

Pediatric plexiform fibromyxoma

A PRISMA-compliant systematic literature review

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Abstract

Background: Plexiform fibromyxoma (PF) is a rare gastric mesenchymal tumor, with approximately 80 cases reported to date. Gastrointestinal stromal tumor, the most common primary mesenchymal tumor of the stomach, shows different biological and clinical characteristics between adult and pediatric patients.

Objectives: This systematic literature review was conducted to elucidate the pathological and clinical features of pediatric PF compared to adult PF.

Methods: MEDLINE (1948 to March 2018) and EMBASE (1947 to March 2018) were searched, and all English articles that reported clinical data on PF patients were identified. Two authors independently reviewed the articles and extracted data to assess immunohistochemistry, sex, chief complaint, tumor size, tumor-related mortality, and tumor recurrence and metastasis.

Results: A total of 41 reports with 80 PF patients (of whom 70 were adult PF and 10 were pediatric PF patients) confirmed by histological and immunohistochemical findings were included. Of a total of 80 tumors, 62 (78%) were located in the gastric antrum, 42 (65%) presented with ulceration, and 48 (74%) were resected by partial gastrectomy. Median tumor size of the resected specimen was larger in pediatric PF than in adult PF cases (5.3 cm vs 4.0 cm, $P = .036$). However, there was no difference between pediatric and adult PFs in immunohistochemical expression, sex predominance, chief complaint, tumor-related mortality, and tumor recurrence and metastasis during the follow-up periods.

Conclusion: Other than increased tumor growth in pediatric PFs, PF is a single disease entity with similar pathological features and benign clinical behavior regardless of onset age.

Abbreviations: GIST = gastrointestinal stromal tumor, PF = plexiform fibromyxoma.

Keywords: adolescent, child, gastrointestinal neoplasms, plexiform angiomyxoid myofibroblastic tumor

What is known

- Plexiform fibromyxoma is a rare mesenchymal tumor presumed to be a benign tumor that most commonly arises in the gastric antrum.
- The most important differential diagnosis with plexiform fibromyxoma is gastrointestinal stromal tumor, in which

the clinical and genetic features differ between pediatric and adult patients.

What is new

- All plexiform fibromyxomas demonstrate benign clinical behaviors, and the tumors grow more in pediatric patients than in adult patients.
- Other than tumor size, there are no pathological or clinical differences between pediatric and adult plexiform fibromyxomas, which indicates a single disease entity irrespective of onset age.

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1. Introduction

Plexiform fibromyxoma (PF) is a rare gastric tumor, officially recognized as a distinct entity among mesenchymal tumors in the 2010 WHO Classification of Tumours of the Digestive System.^[1] After immunohistochemical analyses became available, PF was first described as a plexiform angiomyxoid myofibroblastic tumor of the stomach in 2007.^[2] To date, there have been approximately 80 cases of PF reported.

Gastrointestinal stromal tumor (GIST), which is the most common primary mesenchymal tumor of the stomach, occurs rarely in pediatric patients, and the generally accepted definition of pediatric GIST is based on the diagnosed age of 18 years or

younger.^[3] Pediatric GIST differs from adult GIST with respect to the clinical features of female predominance, gastric location, epithelioid histologic morphology, lower frequency of *KIT* or *PDGFRA* mutations, and an indolent clinical course.^[3] However, the characteristics of pediatric PF remain to be elucidated. This systematic literature review was conducted to clarify the pathological and clinical features of pediatric PF compared to adult PF.

2. Methods

2.1. Criteria for considering articles for review

2.1.1. Types of studies. All types of articles that reported clinical data on PF patients were reviewed, and 1 additional case treated in our center was included in this review. This study protocol was approved by the Beppu Medical Center Ethics Committee.

2.1.2. Types of participants. Adult patients (19 years or older) and pediatric patients (18 years or younger) were diagnosed with PF based on histological examination showing that the tumor tissues exhibited a plexiform intramural growth pattern composed of spindle-shaped bland tumor cells with oval nuclei and abundant myxoid or fibromyxoid stroma with increased vascularity.

Cases reported as PF with potentially different diagnoses, such as GIST, schwannoma, inflammatory myofibroblastic tumor, desmoid-type fibromatosis, and myxoid soft tissue perineuroma, were excluded if the tumor cells were immunohistochemically positive for any of CD34, CD117, discovered on GIST-1 (DOG1), S100, β -catenin, anaplastic lymphoma kinase (ALK), and epithelial membrane antigen (EMA).

2.1.3. Types of outcome measures. In adult and pediatric PF patients separately, the outcomes included: primary outcomes (pathological features), with immunohistochemistry including vimentin, smooth muscle actin (SMA), desmin, caldesmon, muscle specific actin (HHF35), and cytokeratin; and secondary outcomes (clinical features) including sex predominance, chief complaint, tumor size (maximum diameter) of the resected specimen, tumor-related mortality, and tumor recurrence and metastasis during the follow-up period.

2.2. Electronic search methods for identification of articles

MEDLINE (1948 to March 2018) and EMBASE (1947 to March 2018) were both accessed on 13 March 2018, using the following search strategy:

1. Plexiform fibromyxoma [tiab]
2. Plexiform angiomyxoma [tiab]
3. Plexiform angiomyxoid myofibroblastic tumor [tiab]
4. 1 OR 2 OR 3

All English language articles identified were reviewed. Reference lists of the retrieved articles were also searched to find any other relevant published data. There were no date or publication restrictions.

2.3. Data collection and analysis

2.3.1. Article selection and data extraction. Two reviewers (MF, HK) independently reviewed the full text of identified English articles and excluded articles failing to meet the criteria. Data from identified articles that met the criteria was also independently extracted.

2.3.2. Dealing with missing data. Corresponding authors were contacted for missing data where possible, and primary or secondary outcomes were assessed after excluding missing values.

2.3.3. Data analysis. The results from articles that met the criteria were included in the analysis, and the outcomes were assessed separately for adult and pediatric PF cases. Immunohistochemically, diffusely positive or focally positive staining was considered positive expression. Data is expressed as numbers (percentage) for categorical variables and as medians (interquartile range [IQR]) for continuous variables. For statistical analyses, the Chi-squared test, Fisher exact test, and the Mann-Whitney *U* test were used, as applicable. All analyses were performed using SPSS Statistics Version 20 (IBM, Armonk, NY).

3. Results

The electronic search identified 80 records (MEDLINE=43, EMBASE=37). After 23 reports were removed because of duplication, 57 reports were identified for potential inclusion. Full article assessment of 57 reports with the reference search identified 45 reports including 84 cases that met the inclusion criteria. Five cases in 4 reports were excluded because of incompatible immunohistochemistry results, such as positive for CD34 or S100.^[4-7] Including 79 PF cases in 41 reports and 1 PF case treated in our center, a total of 80 PF cases (of which 70 were adult PF and 10 were pediatric PF cases) were included in this review; they are listed in Table 1.^[2,6,8-46] There was complete agreement between the review authors assessing the reports.

Baseline demographic and clinical characteristics of the 80 patients are shown in Table 2. Ulcerative lesions of the tumor were observed in 65% of all patients and in 95% of those with a chief complaint indicating gastrointestinal bleeding, including hematemesis, tarry stool, and anemia. The location of the tumor was in the stomach in 94% of cases, but 6% were in other organs, such as the esophagus, duodenum, jejunum, and cecum.

Immunohistochemically, tumor cells were positive or focally positive for vimentin in 100% of patients examined, smooth muscle actin (SMA) in 93%, desmin in 41%, caldesmon in 57%, muscle-specific actin (HHF35) in 100%, and cytokeratin in 6% (Table 1). In contrast, staining for CD34, CD117, DOG1, S100, neurofilament, β -catenin, ALK, and EMA was negative in all patients examined. Ki-67 proliferation indices were assessed in 14 cases, and those of 13 (93%) were 2% or lower. All 23 cases that were analyzed for *KIT* and *PDGFRA* mutations demonstrated wild-type sequences.

Primary and secondary outcomes were compared between adult and pediatric PF cases (Table 3). Immunohistochemical expressions of pediatric PF were not different from those of adult PF. Median tumor size of the resected specimen was larger in pediatric PF than in adult PF, but sex predominance and chief complaints were not different between the groups. No patients died of PF or had recurrence or metastasis of PF during the follow-up period.

4. Discussion

In summary, both adult and pediatric PF cases showed similar biological and clinical features, except for enlarged tumor size at surgical resection in pediatric PF cases. Regardless of age at onset, PF showed benign clinical behaviors, and no recurrence, metastasis, or tumor-related deaths were observed. This is the

Table 1
Details of 80 cases with immunohistochemical features compatible with PF.

No	Sex	Age	Chief complaint	Tumor location	Ulcer (cm)	size (cm)	Tumor										Immunohistochemistry					Genetic mutation					Follow-up period	References	
							SMA	Vimentin	Desmin	Caldesmon	HHF35	Cyokeratin	CD10	CD34	CD117	D061	S100	Neurofilament	β-catenin	ALK	EMA	Ki-67	KIT	PDGFRα	Treatment				
1	M	50	Abdominal pain	Gastric antrum	Yes	4.0	Pos	Neg	Neg	Focal	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	ND	[2]
2	M	68	Incidental	Gastric antrum	No	4.5	Pos	Neg	Neg	Focal	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	12 months	[2]
3	M	61	Hematemesis	Gastric antrum	Yes	3.7	Pos	Neg	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	6 months	[8]
4	F	19	ND	Gastric antrum	No	4.5	ND	Pos	Focal	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	9 months	[9]	
5	M	46	Gastrointestinal bleeding	Gastrointestinal	Yes	3.5	ND	Pos	Focal	Pos	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	4 months	[9]	
6	F	50	Nausea	Gastric antrum	Yes	1.9	ND	Pos	Focal	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Tumor resection	3 months	[10]	
7	F	7	Vomiting, diarrhea	Gastric antrum	No	15.0	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Tumor resection	ND	[11]	
8	F	16	Hematemesis	Gastric antrum	Yes	10.0	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	3 years	[11]	
9	M	21	Anemia	Gastric antrum	Yes	9.0	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	22 years	[11]	
10	F	30	Intractable gastric ulcer	Gastric antrum	Yes	10.0	Pos (8)	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	9 years	[11]	
11	M	33	Anemia	Gastric antrum	Yes	5.5	ND	Pos	Neg	Neg	ND	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	19.7 years	[11]	
12	F	38	Gastrointestinal bleeding	Gastric antrum	Yes	3.0	ND	ND	Neg (6)	Neg (3)	ND	Neg (3)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Neg (6)	Partial gastrectomy	ND	[11]	
13	M	43	Gastrointestinal bleeding	Gastric antrum	Yes	5.5	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	18.4 years	[11]	
14	M	50	Gastric obstruction	Gastric antrum	No	7.0	Pos	Neg	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	25.5 years	[11]	
15	F	56	ND	Gastric pylorus	No	5.5	Neg (2)	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	19.9 years	[11]	
16	M	62	Weight loss	Gastric antrum	No	4.0	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	ND	[11]	
17	F	65	Weight loss	Gastric antrum	Yes	5.0	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	14.5 years	[11]	
18	F	75	ND	Gastric antrum	Yes	5.0	Pos	Pos	Focal	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	2 months	[12]	
19	F	23	Tarry stool	Gastric antrum	Yes	8.0	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	2 months	[12]	
20	F	54	Abdominal distention	Gastric fundus	No	1.5	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Tumor resection	6 months	[13]	
21	F	35	Incidental	Gastric antrum	No	4.0	ND	Pos	Focal	Pos	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	12 months	[14]	
22	M	34	Abdominal discomfort	Gastric antrum	Yes	3.5	ND	Pos	Focal	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	2 months	[15]	
23	M	23	Abdominal pain, tarry stool	Gastric antrum	ND	14.0	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	12 months	[16]	
24	F	44	ND	Cecum	No	5.0	Pos	Neg	Neg	ND	ND	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	ND	ND	[17]	
25	M	52	Abdominal discomfort	Gastric antrum	Yes	3.5	ND	Pos	Focal	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Tumor resection	5 months	[18]	
26	M	47	Incidental	Gastric body	Yes	3.0	ND	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Tumor resection	6 years	[19]	
27	F	63	Incidental	Gastric body	Yes	2.2	ND	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Tumor resection	ND	[19]	
28	F	42	Abdominal pain	Gastric antrum	Yes	12.9	Pos	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	3 weeks	[20]	
29	F	27	Abdominal pain, tarry stool	Gastric antrum	Yes	4.6	ND	Pos	Focal	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	40 months	[21]	
30	M	32	Incidental	Gastric antrum	No	3.4	Pos	Focal	Focal	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	3 years	[22]	
31	M	60	Abdominal pain	Gastric antrum	No	2.0	ND	Neg	ND	ND	ND	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	12 months	[23]	
32	F	11	Anemia	Gastric pylorus	Yes	3.5	Pos	Pos	Pos	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	15 months	[24]	
33	F	16	Incidental	Esophagus	No	3.2	ND	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Tumor resection	14 months	[24]	
34	F	38	Incidental	Gastric body	Yes	3.5	ND	Pos	ND	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Tumor resection	6 months	[25]	
35	F	19	Abdominal pain	Duodenum	Yes	13.8	ND	Neg	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Duodenectomy	6 months	[26]	
36	F	26	Vomiting, tarry stool	Gastric antrum	Yes	ND	Pos	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	ND	[27]	
37	F	40	Abdominal discomfort	Gastric antrum	No	9.5	Pos	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	ND	[28]	
38	F	36	Abdominal pain, anemia	Gastric antrum	Yes	2.5	ND	ND	ND	ND	ND	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Tumor resection	ND	[29]	
39	M	39	Abdominal pain	Gastric antrum	No	3.5	ND	Pos	ND	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	9 months	[30]	
40	F	28	Abdominal pain, anemia	Gastric antrum	Yes	5.5	ND	Pos	ND	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	23 months	[31]	
41	M	51	Abdominal pain	Gastric antrum	Yes	7.0	Pos	Pos	Neg	Neg	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	ND	[32]	
42	M	47	Vomiting	Gastric antrum	Yes	6.0	Pos	Pos	Focal	Focal	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	10 months	[33]	
32	F	28	Abdominal pain	Gastric antrum	Yes	3.0	ND	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Partial gastrectomy	ND	[34]	
44	F	18	ND	Gastric antrum	ND	4.5	ND	Focal	ND	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	ND	ND	[35]	
45	M	19	ND	Gastric antrum	ND	5.5	ND	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	ND	ND	[36]	
46	F	19	ND	Stomach	ND	4.5	ND	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	ND	ND	[36]	
47	M	29	ND	Gastric antrum	ND	6.5	ND	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	ND	ND	[36]	
48	F	30	ND	Stomach	ND	2.0	ND	Pos	Pos	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	ND	ND	[36]	
49	F	36	ND	Gastric antrum	ND	8.0	ND	Pos	Neg	ND	Pos	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	Neg	ND	ND	[36]	

(continued)

Table 1
(continued).

No.	Sex	Age	Chief complaint	Tumor location	Ulcer (cm)	size (cm)	Tumor										Immunohistochemistry					Genetic mutation					Follow-up period	References								
							Vimentin	SMA	Desmin	Caldesmon	HHF35	Cytokeratin	CD10	CD34	CD117	DOG1	S100	Neurofilament	β-catenin	ALK	EMA	Ki-67	KIT	PDGFRA	Treatment											
50	M	46	ND	Gastric antrum	ND	3.5	ND	Pos	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[6]
51	F	47	ND	Gastric antrum	ND	4.5	ND	Pos	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[6]
52	F	51	ND	Gastric antrum	ND	9.0	ND	Pos	Focal	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[6]	
53	F	58	ND	Gastric antrum	ND	ND	ND	Pos	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[6]	
54	F	62	ND	Stomach	ND	ND	ND	Pos	Focal	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[6]		
55	F	63	ND	Jejunum	ND	3.5	ND	Pos	Focal	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[6]		
56	F	65	ND	Gastric antrum	ND	4.3	ND	Pos	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[6]	
57	F	76	ND	Stomach	ND	4.0	ND	Focal	Pos	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[6]		
58	F	9	Abdominal pain, vomiting	Gastric antrum	Yes	5.0	ND	Pos	Focal	ND	ND	Focal	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[59]		
59	M	55	Incidental	Gastric antrum	No	1.7	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[97]		
60	M	11	Abdominal pain	Gastric pylorus	No	17.0	ND	Focal	Neg	Focal	ND	ND	Neg	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[97]		
61	M	63	Anemia, tarry stool	Duodenum	Yes	3.0	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[98]		
62	M	51	Abdominal discomfort	Gastric antrum	No	4.0	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[98]		
63	M	66	ND	Gastric antrum	No	4.0	ND	Focal	Neg	Focal	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[40]		
64	F	16	Anemia	Gastric antrum	Yes	6.5	ND	Pos	Focal	ND	ND	Focal	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[41]		
65	M	34	Abdominal discomfort	Gastric antrum	No	1.6	ND	Pos	Neg	ND	ND	Focal	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[41]		
66	M	51	Incidental	Gastric antrum	No	1.9	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[42]		
67	F	41	Anemia, tarry stool	Gastric antrum	Yes	5.5	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[43]		
68	M	26	Abdominal distention	Gastric fundus	Yes	1.8	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[44]		
69	F	31	Abdominal distention	Gastric antrum	No	2.5	ND	Pos (10)	Focal (6)	ND	ND	Focal (5)	ND	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	ND	[44]		
70	F	40	Tarry stool	Gastric antrum	Yes	3.8	ND	Focal (6)	Focal (6)	ND	ND	Focal (5)	ND	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	[44]		
71	M	46	Abdominal distention	Gastric antrum	No	4.0	ND	Pos (10)	Neg (5)	ND	ND	Neg (5)	ND	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	[44]		
72	M	48	Abdominal distention	Gastric antrum	Yes	3.0	Pos (10)	Pos (10)	Neg (5)	ND	ND	Neg (5)	ND	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	[44]		
73	M	52	Incidental	Gastric fundus	Yes	1.2	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[44]	
74	M	58	Abdominal pain	Gastric antrum	Yes	3.5	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[44]	
75	F	59	Abdominal distention	Gastric antrum	Yes	2.5	ND	Pos (10)	Neg (4)	ND	ND	Neg (5)	ND	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	Neg (10)	[44]		
76	F	59	Tarry stool	Gastric antrum	Yes	3.0	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[44]	
77	F	72	Abdominal distention	Gastric antrum	Yes	7.0	ND	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[44]	
78	M	14	Anemia	Gastric antrum	No	5.0	ND	Pos	Focal	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[49]	
79	M	37	Hematemesis	Gastric antrum	Yes	5.8	ND	Pos	Pos	ND	ND	Pos	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	[49]	
80	F	14	Abdominal pain, hematemesis (unpublished)	Gastric antrum	Yes	5.5	Pos	Pos	Neg	ND	ND	Neg	ND	Neg	ND	ND	ND	ND	ND	Neg	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	ND	Present case	

ALK = anaplastic lymphoma kinase, DOG-1 = discovered on GIST-1, EMA = epithelial membrane antigen, Focal = focally positive, HHF35 = muscle-specific actin, ND = not described, Neg = negative, Pos = positive, SMA = smooth muscle actin.

Table 2**Baseline demographic and clinical characteristics.**

	Plexiform fibromyxoma (n=80)
Female, n (%)	46 (58)
Age (y)	42 (27–55)
Chief complaint, n (%)	
Abdominal pain	15 (25)
Incidental	10 (16)
Anemia	9 (15)
Tarry stool	8 (13)
Abdominal distention	7 (11)
Abdominal discomfort	5 (8.2)
Hematemesis	4 (6.6)
Vomiting	4 (6.6)
Gastrointestinal bleeding	3 (4.9)
Weight loss	2 (3.3)
Others	4 (6.6)
Tumor location, n (%)	
Gastric antrum	62 (78)
Other gastric sites	13 (16)
Duodenum	2 (2.5)
Esophagus	1 (1.3)
Jejunum	1 (1.3)
Cecum	1 (1.3)
Ulceration of tumor, n (%)	42 (65)
Tumor size (cm)	4.0 (3.1–5.7)
Treatment, n (%)	
Partial gastrectomy	48 (74)
Tumor resection	15 (23)
Duodenectomy	2 (3.1)
Follow-up period (months)	16 (6.8–53)

Data is frequencies (%) or medians (interquartile range).

first systematic literature review to assess the age-related characteristics of PF.

The annual incidence rate of GIST was reported to be 0.68 per 100,000 in the United States.^[47] Although the epidemiology of PF remains unclear, the incidence of PF is estimated to be less than 1/150 of that of GIST,^[11] and the reported frequency of adult PF was seven times higher than of pediatric PF in the current study.

Based on these data, the estimated annual incidence rate of PF is approximately 0.45 (0.40 in adult and 0.05 in pediatric PF) per 100,000,000. Thus, PF is a rare submucosal tumor, but it should be considered as a potential cause of mesenchymal tumors of the stomach, especially in the antrum.

The clinical features of pediatric GIST are similar to those of pediatric PF, characterized by commonly arising from the gastric antrum, exhibiting myxoid morphology, and the absence of *KIT* or *PDGFRA* mutations.^[3] In contrast, pediatric GIST can be immunohistologically discriminated from pediatric PF by sensitive and specific immunohistochemical markers, such as CD34, CD117, and DOG1.^[48] Similarly, several gastrointestinal tumors with histology similar to pediatric PF can be discriminated by immunohistochemical markers, such as CD34 for inflammatory fibroid polyp, S100 for gastrointestinal schwannoma, β -catenin for desmoid-type fibromatosis, ALK for inflammatory myofibroblastic tumor, and EMA for myxoid soft tissue perineuroma.^[48] Based on these pathological findings, the current study excluded atypical PFs with incompatible immunohistochemistry and included immunohistochemically compatible PFs, and the pathological and clinical features of pediatric PF were elucidated. The immunohistochemistry of pediatric PF was similar to that of adult PF with respect to positive expressions of specific markers, such as of mesenchymal cells (vimentin), muscular cells (SMA, desmin, caldesmon, and HHF35), and epithelial cells (cytokeratin). In addition, there were common clinical characteristics, such as hemorrhagic ulcer formation and a benign clinical course, shared by pediatric and adult PF cases. Therefore, the results from the present study are biologically plausible with respect to pathological and clinical compatibility. The enlarged tumor size of pediatric PF could be explained by age-related higher tumor cell proliferation and might be associated with a lower frequency of incidental diagnosis in pediatric PF than in adult PF cases. The present study showed similar pathological and clinical characteristics between pediatric and adult PF cases, unlike pediatric and adult GIST cases, and this finding suggests that PF is a single disease entity unrelated to the age at onset. PF can be treated and followed-up as a benign tumor when confirmed histologically and immunohistochemically.

Table 3**Primary and secondary outcome measures.**

	Adult PF	Pediatric PF	Risk ratio (95%CI)	P value
	≥19 years (N=70)	≤18 years (N=10)		
Positive immunohistochemistry, n (%)				
Vimentin	21 (100)	2 (100)	–	–
SMA	55 (95)	8 (100)	1.1 (1.0 – 1.1)	.67
Desmin	21 (42)	5 (63)	1.5 (0.8 – 2.8)	.24
Caldesmon	12 (63)	2 (100)	1.6 (1.1 – 2.2)	.43
HHF35	10 (100)	–	–	–
Cytokeratin	1 (8.3)	0 (0.0)	0.9 (0.8 – 1.1)	.86
Female, n (%)	38 (54)	8 (80)	1.5 (1.0 – 2.1)	.11
Chief complaint, n (%)				
GI bleeding-associated symptom	17 (33)	5 (56)	1.7 (0.8 – 3.4)	.17
Incidental	9 (17)	1 (11)	0.6 (0.1 – 4.5)	.54
Tumor size (cm)	4.0 (3.0 – 5.5)	5.3 (4.3 – 11)	–	.036
Died of PF, n (%)	0 (0)	0 (0)	–	–
Recurrence, n (%)	0 (0)	0 (0)	–	–
Metastasis, n (%)	0 (0)	0 (0)	–	–
Follow-up period (months)	18 (6.8 – 67)	15 (7.5 – 31)	–	.63

Data is frequencies (%) or medians (interquartile range).

GI bleeding-associated symptom means anemia, gastrointestinal bleeding, hematemesis, or tarry stool.

GI=gastrointestinal, HHF35=muscle-specific actin, PF=plexiform fibromyxoma, SMA=smooth muscle actin.

This study had some limitations because of the literature review and small sample size. In view of potential unreported PF cases without severe symptoms, there might be a risk of selection bias toward overestimation of severity. However, under such conditions, there were no tumor-related deaths, recurrences, or metastases of PF, which provides the basis for considering PF a benign tumor. A population-based registry for PF is required to further investigate the epidemiology of PF.

In conclusion, both pediatric and adult PF cases showed similar pathological characteristics and benign clinical behavior, which indicates that PF is the same disease entity, regardless of onset age.

Author contributions

MF and HK wrote the manuscript. MF and HK reviewed the literature. SH, TS, YN, and YY assessed the clinical data, histology, and immunohistochemical findings of included cases. MF and HK developed the concept of this study. All authors commented on the drafts.

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