CASE REPORT

Infantile fibrosarcoma of ethmoid sinus, misdiagnosed as an adenoid in a 5-year-old child

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ABSTRACT

Infantile fibrosarcoma of head and neck is rare and the presence of this tumor in ethmoid sinus is even more uncommon. To the best of our knowledge, <5 cases have been reported in the last 20 years in the English literature, so far, only one of which has been infantile type in a 15 months old girl. In this case report, we will explain our experience with a rare case of infantile fibrosarcoma originating from ethmoid sinus in a 5-year-old boy who presented with dyspnea and epistaxis. After biopsy, it was diagnosed as fibrosarcoma of sinus origin.

Key words: Ethmoid sinus, epistaxis, infantile fibrosarcoma

INTRODUCTION

Fibrosarcoma is a common soft tissue sarcoma and constitutes more than 6% of sarcomas in the whole body, most commonly occurs and originates from the extremities, however its occurrence in head and neck is not common accounting for <1% of the malignancies in this area.[1]

Fibrosarcoma in the pediatric age group is also rare; originating most commonly from the soft tissue of extremities, its occurrence in the sinus area, especially ethmoid sinus is an extremely rare event.[2]

In this report, we present a case of a 5-year-old boy who presented with dyspnea and nose bleeding, with the clinical diagnosis of adenoid. However after biopsy, the pathologic diagnosis was fibrosarcoma of ethmoid sinus.

CASE REPORT

A 5-year-old boy presented with nose bleeding and dyspnea of 2-month duration. He has been completely well, with no positive past medical history.

His parents have noticed significant snoring at sleep since 7 months prior admission. At that time when they referred to their family physician, they were reassured that their son has an adenoid. He also had decreased appetite during the last 2 months.

He has been the first child of the family, born with normal vaginal delivery.

Physical examination showed a healthy boy with loud breathing sound.

Heart rate: 95/min, respiratory rate: 15/min, temperature: 37°C and blood pressure were normal.

His laboratory data was normal, complete blood count, blood urea nitrogen/creatinine, liver function tests and fasting blood sugar were all normal.

After worsening of the boy's dyspnea and nose bleeding, a computed tomography scan of head and neck was ordered which showed:

A large enhancing lesion in both ethmoidal sinuses and both nasal cavities with extension to nasopharynx, causing near total obliteration of the nasopharynx was noticed. This lesion

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showed heterogeneous enhancement with the erosion of bony parts of paranasal sinuses. The lesion had caused septal deviation to the right side [Figure 1a and b].

The patient was operated to take a biopsy for primary diagnosis and to take a decision on further treatment. Pathologic sections showed a moderately cellular tumor with spindle shaped cells the cells showed typical herring-bone pattern [Figure 2a-d]. Individual tumor cells were spindle shaped with mild to moderate atypia and indistinct cytoplasmic borders. There was no significant necrosis and mitotic figures were <2/10 HPF. Immunohistochemistry was positive for vimentin and negative for desmin, smooth muscle actin (SMA), MyoD1, CD31, neuron specific enolase (NSE), S100, synaptophysin, CD56, minimum inhibitory concentration-2 (MIC-2) and MIB-1 proliferative index was < 5% [Figure 2e].

The tumor cells were negative for SMA and desmin, so leiomyosarcoma was excluded and the tumor cells were negative for neurogenic markers like CD 56, S-100 and NSE, hence nerve sheath tumors were excluded. Due to the typical histology, presence of herring-bone pattern and immunohistochemical findings i.e., positivity only for vimentin, the diagnosis of low grade fibrosarcoma was made and the patient was scheduled for operation by an ear-nose-throat surgeon.

Endoscopic nasal evaluation revealed a large mass to be originating from the anterior and a part of posterior ethmoid cells with extension to nasopharynx, without nasopharyngeal involvement. The mass was completely resected after anterior ethmoidectomy. Then frontal maxillary sinus which was filled with mucopurulent material was drained. The mucosa was inflamed. Multiple biopsies were taken from frontal recess to evaluate its involvement, but maxillary ostium did not show any tumoral involvement.

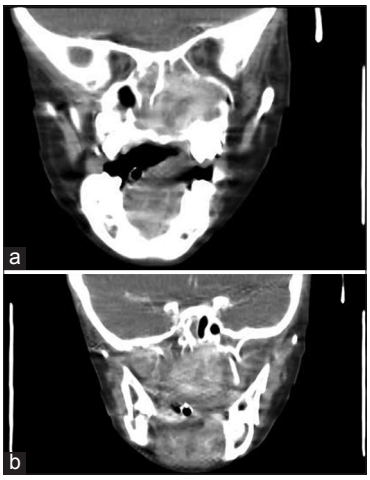

Figure 1: (a) Computed tomography image shows the lesion, causing septal deviation to the right side. (b) Computed tomography image with the lesion showing heterogeneous enhancement and erosion of the bony parts of paranasal sinuses

The pathology specimen received in the laboratory showed the same pattern of the biopsy with moderate cellularity and spindle-shaped cells with herring-bone pattern. The additional biopsies from maxillary ostium and frontal recess were free.

There has been no tumor residue after the surgery, so no adjuvant treatment was prescribed.

After 6 months, the patient was reevaluated, he is doing well and all of the imaging studies done are negative, that is, no residual tumor tissue has been identifi ed. The decision was made not to prescribe any adjuvant treatment. The patient will be under strict follow-up for the early detection of recurrence.

DISCUSSION

Soft tissue sarcomas constitute 5–15% of malignancies in the pediatric age group and only 5% of the sarcomas in this age group are located in the head and neck region.[3]

The most common sarcoma in children in the head and neck area is rhabdomyosarcoma.[4]

Fibrosarcoma is a rare sarcoma in children accounting for <10% of this type of malignancies in the pediatric age group.[5] Its most common location in children is the extremities. Head and neck is the second common location in this age group (<1% of fibrosarcomas).[6]

It is observed that that pediatric fibrosarcoma shows two peaks of age presentation, one in early infancy (most studies believe <1-year of age) and the other in the older children.[7]

Previous literature did not show any definite opinion regarding the exact age for use of the term infantile fibrosarcoma, however some authors have mentioned a cut-off age of 2 years, and others have suggested a cut-off age of up to 5 years.[8]

Clinical behavior of infantile fibrosarcoma during the first 5 years of age differs from that observed in the older children and adults.[9]

In this case report, we describe our experience with a 5-year-old boy with fibrosarcoma of ethmoid sinus, diagnosed after pathologic examination of the surgical biopsy. He presented with nose bleeding, dyspnea and snoring for about 2 months.

Clinical manifestations of soft tissue sarcomas in the head and neck are different and depend on the site of tumor; those in the area of nasopharynx, sinuses and nose, mostly have been reported to present with nasal obstruction and epistaxis.[10]

The only way of diagnosis is by pathologic examination after imaging studies. Histologically, sinonasal fibrosarcomas are composed of hypercellular tumors with spindle shaped thin

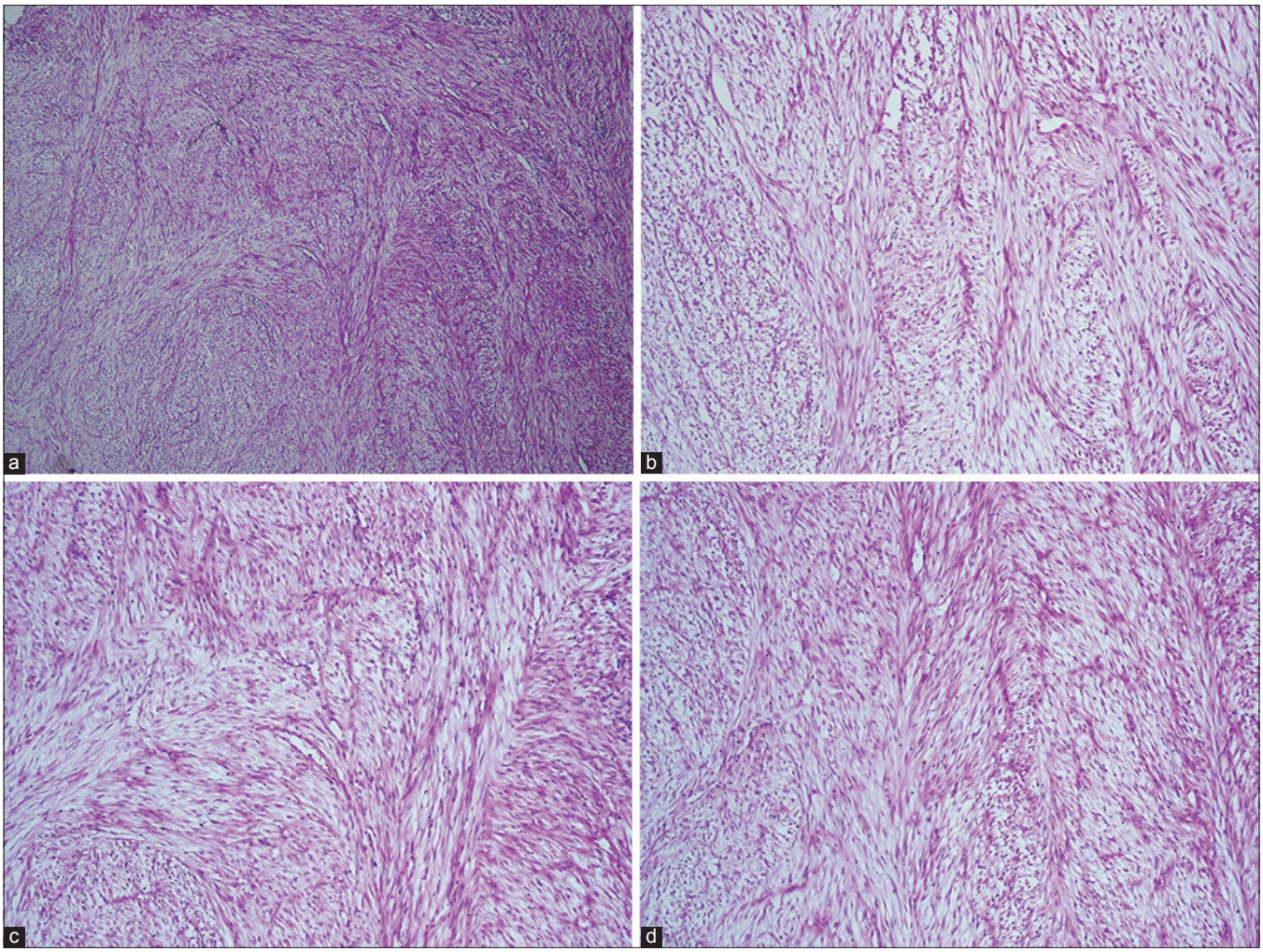

Figure 2: : (a) Pathologic sections showed moderately cellular tumor with spindle shape cells (H&E stain, ×100). (b) Sections from the tumor showing typical herring bone pattern (H&E stain, ×250). (c) Sections show spindle-shaped cells with minimal atypia, low mitosis and herring bone pattern (H&E stain, ×250). (d) High power view shows spindle shaped cells with indistinct cytoplasmic borders (H&E stain, x250).

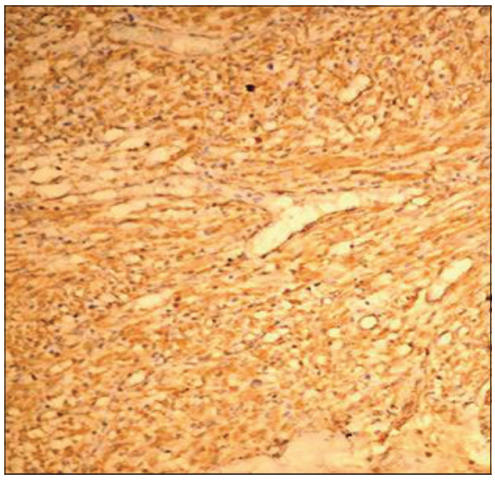

Figure 2e: The tumor cells were positive for vimentin (IHC stain, x100).

cells arranged in typical herring bone pattern.[10] When this histologic picture is diffuse and uniform, diagnosis can be made even without ancillary studies.[3]

These tumors are negative for pancytokeratin, desmin, S100 and SMA. They are only positive only for vimentin.[7]

The treatment of choice is wide surgical excision. Prognosis is excellent with complete resection, although rare tumor recurrence has been reported.[9]

Complete surgical resection is usually curative but chemotherapy may shrink the tumor to facilitate complete resection.[11]

Adjuvant chemotherapy or radiotherapy have also been used in cases in which there have been positive surgical margins or in cases where macroscopically incomplete excision has been carried out.[12] In our case, complete excision was accomplished after surgery with no tumor residue.

Primary fibrosarcoma of ethmoid is extremely rare and to the best of our knowledge, <5 cases have been reported in the English literature so far.[12-16] However, only one case of

infantile fibrosarcoma in the ethmoid sinus has been reported in the literature in a 15-month-old girl.[16] Both of these cases have been treated with antibiotics for a few weeks with no response.

On conclusion, in a patient with prolonged symptoms of nasal obstruction and no response to antibiotics, diagnosis of this rare tumor by tissue biopsy should be considered so that there is no delay in the treatment.

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Conflicts of interest

There are no conflicts of interest.

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