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Secondary Aneurysmal Bone Cyst in a Craniofacial Fibrous Dysplasia: Case Report

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Aneurysmal bone cyst (ABC) is a rare non-neoplastic bone lesion that involves mostly the long bones and vertebrae and may occur very rarely in the craniofacial bones. ABCs may occur as secondary bony pathologies in association with various benign and malignant bone tumors and with fibrous dysplasia (FD). FD is a common non-neoplastic bony pathology mostly affecting craniofacial bones. Secondary ABC occurring in craniofacial FD is extremely rare, with only approximately 20 cases reported in the literature to date. Here, we report on a case of secondary ABC in a 25-year-old woman who has had a craniofacial deformity for over 10 years and who presented to us with a rapidly growing painful pulsatile mass in the right frontal region that began over 2 months prior to admission. On thorough examination of computed tomography and magnetic resonance imaging brain scans taken at two-month interval, an aggressive, rapidly enlarging ABC, arising from the right frontal FD, was diagnosed. The patient underwent preoperative embolization followed by gross total resection of the ABC and cranioplasty. The 6-month follow up showed no recurrence of the ABC, nor was any progression of the FD noticed.

Key Words Aneurysmal bone cysts; Fibrous dysplasia of bone; Frontal bone; Craniotomy.

INTRODUCTION

Aneurysmal bone cyst (ABC), a term now regarded as a misnomer, is a non-neoplastic bony lesion that consists of cystic cavernous cavities without lining endothelium filled with blood and that occurs commonly in teenagers or young adults. ABCs occur mostly in long bones and the spinal column, and craniofacial involvement is a very rare occurrence. ABCs are either primary or secondary; secondary ABCs develop in preexisting bony pathologies primarily including giant cell tumor, osteoblastoma, angioma and chondroblastoma and less commonly including fibrous dysplasia (FD), ossifying fibroma and osteosarcoma.

FD is also a rare pathology that represents 2.5% of all bone tumors and 7% of benign bone tumors [1] and occurs mostly in the craniofacial skeleton of adolescent and young adult patients. FDs are benign non-neoplastic lesions commonly presenting with craniofacial disfigurement or compressive cranial neuropathies such as vision loss. A few cases have been documented with oculomotor palsy due to involvements of the neural foramina at the base of the skull. Most FDs are self-limiting, with stabilization after puberty.

Reports of secondary ABC occurring in craniofacial FDs are extremely rare in the literature, accounting for less than 20 cases. Patients usually present with a rapidly enlarging, soft, painful mass in the preexisting FDs, for which treatment is a curative resection of the ABC with a conservative resection of the FD or a conservative treatment that includes percutaneous sclerosing therapy. The case review presented here was conducted with the approval of the Institutional Review Board (KUH10700028).

CASE REPORT

A 25-year-old female, with no significant medical or familial history, presented with rapidly growing right forehead mass beginning 2 months prior to admission. She had been followed for a diffuse forehead bulging since early in her second decade

of life at a neurosurgical center in Kazakhstan. She underwent a magnetic resonance imaging (MRI) scan to assess the soft bulging in her right frontal region 2 months prior to admission. The mass grew rapidly and she began to feel pain when the mass reached the size of a tennis ball. The patient was transferred to our hospital for definite diagnosis and treatment. The physical examination was normal except for a bulging, soft mass measuring 5.5 cm on the right forehead. She was free of neurological symptoms, and routine laboratory examinations were within normal limits. A computed tomography (CT) scan

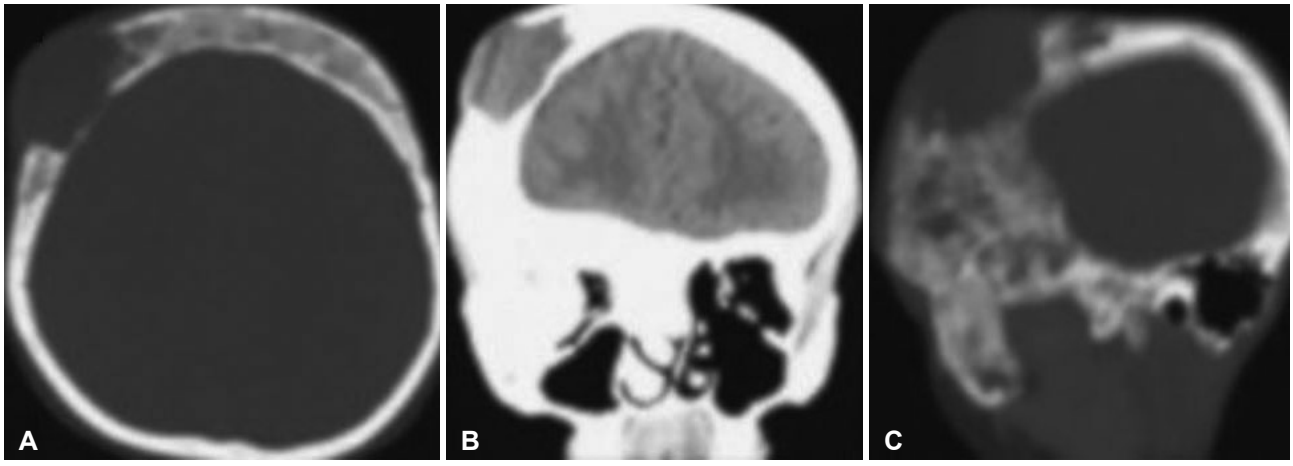

Fig. 1. Preoperative CT scan of the patient; axial (A), coronal (B), and sagittal (C) image. Image shows diffuse fibrous dysplasia in the frontal bone with an osteolytic lesion.

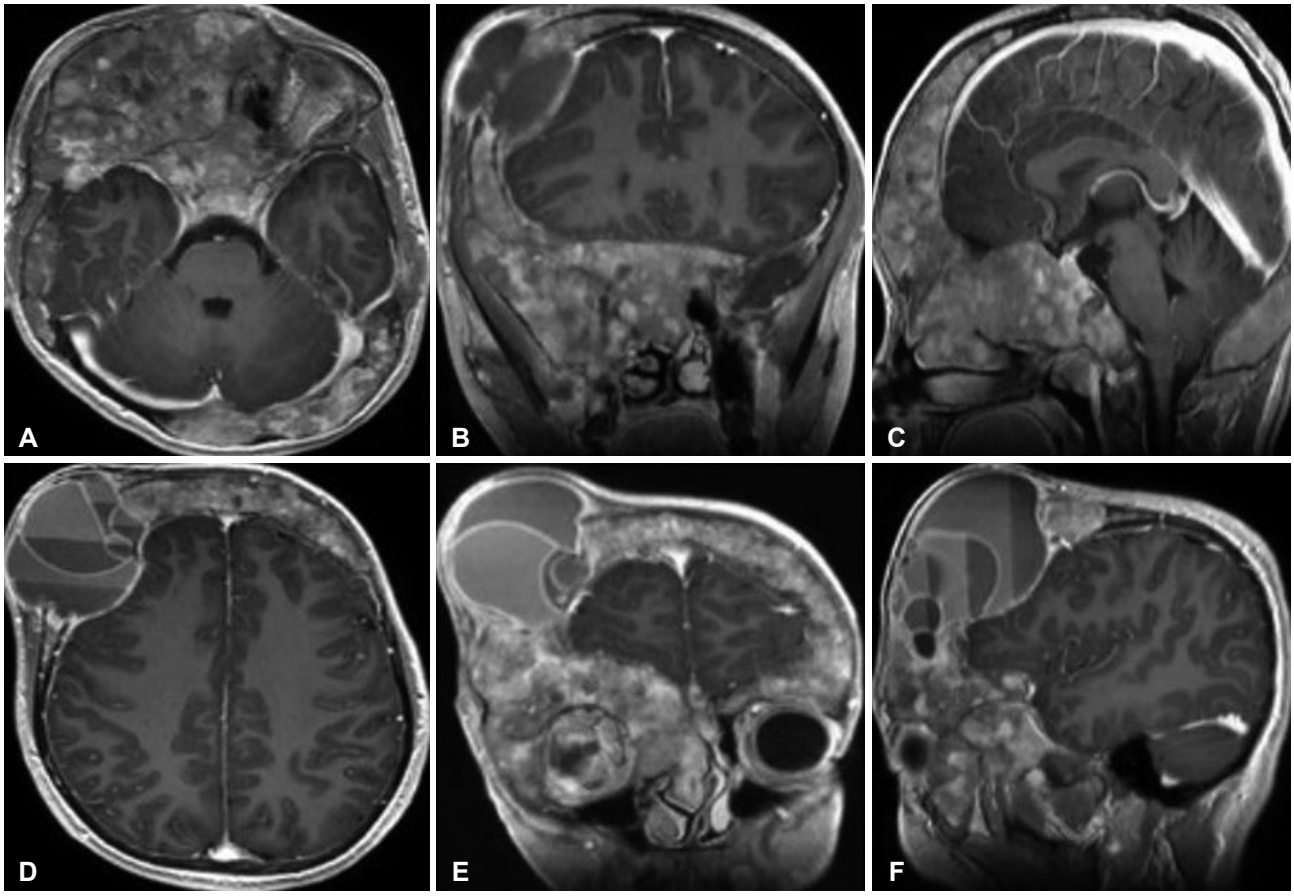

Fig. 2. MRI, gadolinium-enhanced T1-weighted image. Extensive fibrous dysplasia involving frontal, sphenoid, and ethmoid bones, midline parietal bones, both temporal bones, midline and left occipital bone, clivus, and right zygomatic bone (A: axial image, B: coronal image, C: sagittal image). Fluid-fluid level with a large aneurysmal bone cyst (6.2×5.5×5.0 cm) was seen in the right frontal bone (D: axial image, E: coronal image, F: sagittal image).

revealed characteristic findings of FD with a heterogeneous ground-glass appearance of both the frontal, sphenoid, and ethmoid bones, as well as a discrete cystic mass in the right frontal bone (Fig. 1). MRI also showed findings of FD in the bilateral craniofacial bone and indicated multi-septated fluid-fluid levels suggestive of ABC (Fig. 2). We performed a preoperative ophthalmological evaluation, in which visual acuity was 1.0 in both eyes, and there were no visual field, fundus, or eye movement abnormalities. For evaluation of the vascularity of the cystic mass and embolization of feeder arteries prior to surgery, a trans-femoral cerebral angiography was done. The angiography demonstrated moderate vascularity along the rim of the cystic mass and embolization via the right middle meningeal artery and right superficial temporal artery was performed. After embolization, the vascularity of the mass was markedly reduced (Fig. 3).

The patient underwent right frontal craniectomy with gross total resection of the brownish soft-tissue mass with multiple septated, brownish fluids and sediment-filled, cystic cavities followed by simple cranioplasty. The outer table thinned out as fine as paper, and the inner table was nearly absent. The histopathological findings of the skull lesion indicated FD with degenerative changes, forming typical ABC-like areas (Fig. 4). There was no evidence of malignancy in the surgical specimen. The FD was partially removed around the ABC, and after surgery (Fig. 5), the patient's cranio-facial deformity was dramatically improved (Fig. 6). No recurrence of the ABC nor progression of the FD was found at 6 months postoperatively.

DISCUSSION

FD, Langhans cell histiocytosis (Histiocytosis X), and ABC are the three most common juvenile craniofacial bony lesions. Among these, FD is the most common proliferating craniofacial bony lesion. In FD, abnormal growth of fibroblasts replaces medullary bone with fibrocellular tissue [2,3]. The biologic behavior of FD is known to be self-limiting, with no progression after puberty [4-6]. According to reported cases, in FD patient, presented with painful swelling over a short period of time, which may clinically represent the very unusual sarcomatous transformation of FD [7].

ABC is neither an aneurysm nor a true cyst but is made of channels containing flowing blood. The pathogenetic mechanism of ABC remains unclear. Other report suggested that it may arise from abnormal hemodynamic conditions causing an increase in venous pressure, which then leads to hemorrhage

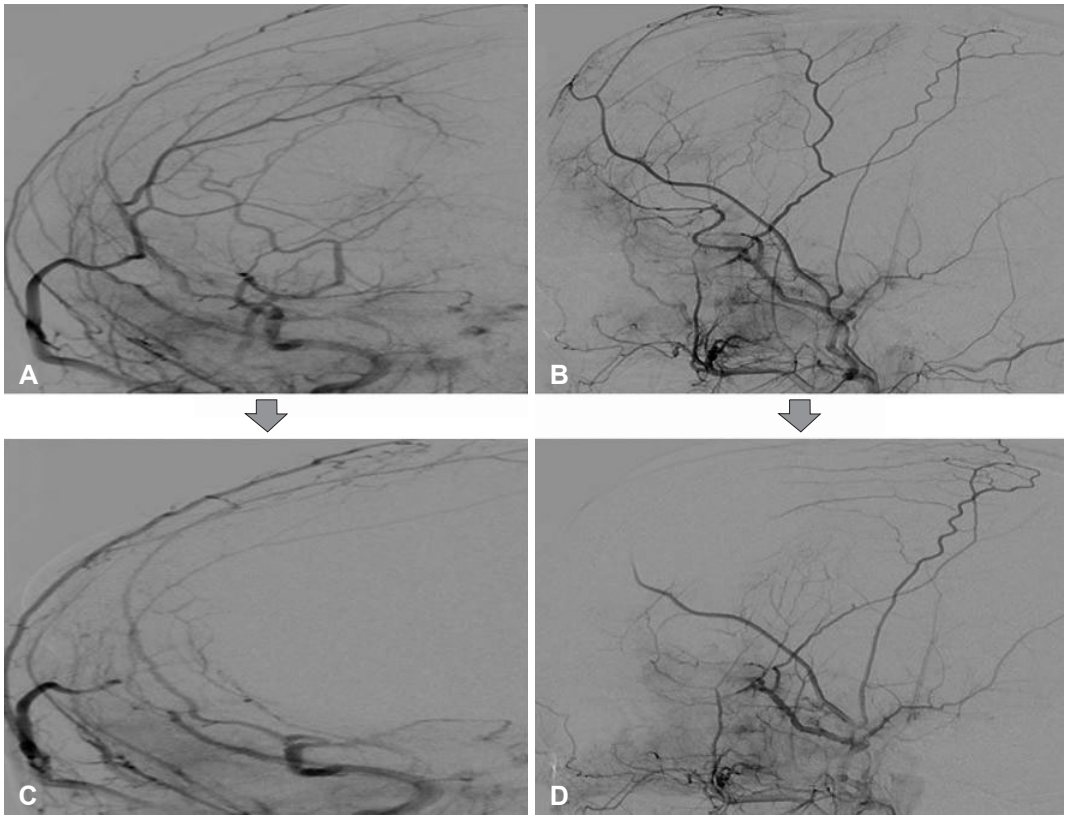

Fig. 3. Trans-femoral cerebral angiography image of the patient. The pre-embolization image showed prominent vascular stain in the right frontal area, at the margin of the aneurysmal bone cyst (A: anterior-posterior view, B: right lateral view). An endovascular embolization was performed in a branch of the right middle meningeal artery and the right superficial temporal artery. After embolization, vascular staining was markedly decreased, but residual staining was seen in the posterior and inferior margins (C: anterior-posterior view, D: right lateral view).

[8]. ABC may occur as either a primary (70%) or a secondary (30%) bone cyst [9]. Diseases known for predisposing the patient to lesions of ABC include osteoclastoma, osteosarcoma, osteoblastoma, and hemangioma [10]. ABC arising from FD is extremely rare, especially in the craniofacial bone. This lesion was first reported by Branch et al. [10] in 1986, and fewer than twenty cases have been reported since then [3,9-21]. We summarized the case reports of ABC with FD in the craniofacial

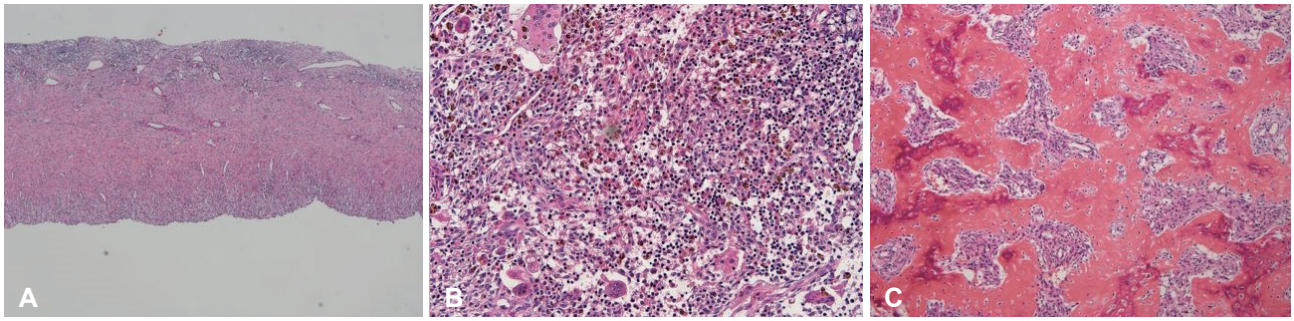

Fig. 4. Histological examination of the resected lesion. A: The aneurysmal bone cyst wall (hematoxylin and eosin staining, original magnification ×100). B: A multinucleated osteoclastic giant cell (hematoxylin and eosin staining, original magnification ×200). C: Woven bone with proliferating fibrous tissue (hematoxylin and eosin staining, original magnification ×200).

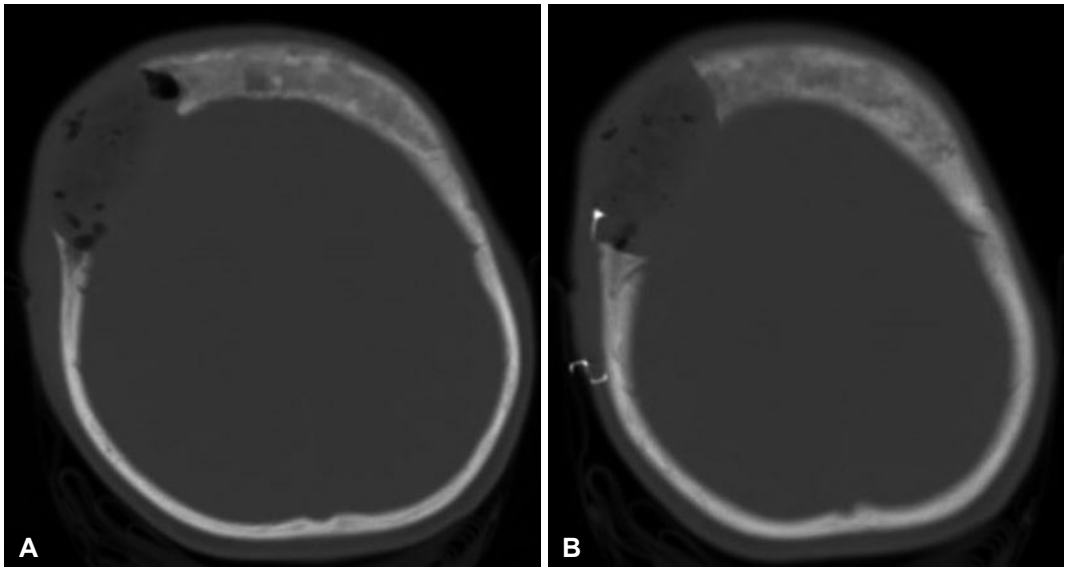

Fig. 5. Postoperative axial CT scan of the setting bone shows the partial removal of the fibrous dysplasia around the aneurysmal bone cysts (A and B).

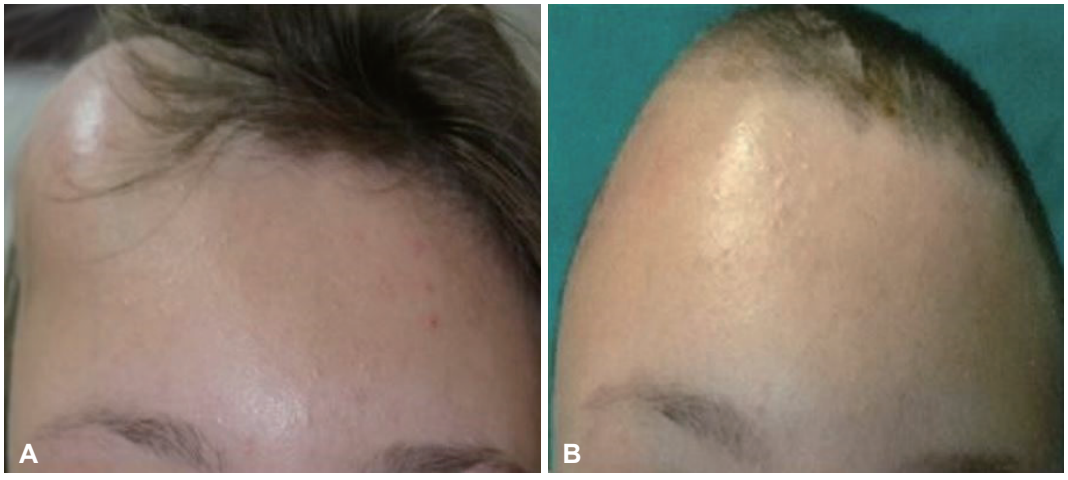

Fig. 6. Cosmetic improvement of the patient's forehead (A: preoperative image, B: postoperative image).

Table 1. Published cases of secondary aneurysmal bone cyst with craniofacial fibrous dysplasia

Authors	Years	Sex	Age	Site	Presenting symptom	Treatment
Branch et al. [10]	1986	M	19	Parietal	Growing mass	Surgical resection
		F	9	Parietal, frontotemporal	Painful mass	Surgical resection
Rappaport [11]	1989	M	25	Occipital	Painful mass	Surgical resection
Wojno and McCathy [12]	1994	F	14	Temporal	Painless mass	Surgical resection
		M	40	Frontal	Exophthalmos and diplopia	Surgical resection
Haddad et al. [13]	1998	M	6	Temporal	Growing mass	Surgical resection
Saito et al. [14]	1998	M	11	Nasal cavity and skull base	Nasal obstruction and headache	Surgical resection
Itshayek et al. [15]	2002	M	19	Occipital and clivus	Painless mass	Endovascular embolization and surgical resection
Pasquini et al. [16]	2002	M	5	Skull base	Sinusitis	Endoscopic trans-nasal resection
Lin et al. [9]	2004	M	18	Frontal	Severe headache	Surgical resection
Iseri et al. [17]	2005	F	35	Occipital and clivus	Severe headache	Medical treatment*
Mattei et al. [3]	2005	M	19	Occipital	Severe headache and nuchal rigidity	Surgical resection
Lee et al. [18]	2010	F	18	Fronto-parietal	Growing mass	Surgical resection
Terkawi et al. [19]	2011	F	7	Nasal cavity and skull base	Vision loss and nasal obstruction	Surgical resection
Manjila et al. [20]	2013	M	10	Nasal cavity and skull base	Cognitive impairment	Surgical resection and endoscopic resection
Hnenny et al. [21]	2015	F	28	Nasal cavity and skull base	Vision worsening	Surgical resection

*Patient refused surgery, medical treatment was done with bisphosphonate

bone in a literature search from PubMed in Table 1. In our patient and in seven of the reported cases, a growing craniofacial mass was a presenting symptom. The rapidly distended contour of a secondary ABC may create suspicion of a malignant change in the pre-existing, benign bone tumor. The incidence of malignant transformation in FD has been reported to be between 0.5% and 4% [7]. When a patient presents with a rapid growth of the skull, the possibility of malignant change of the benign bone pathology and the development of secondary ABC should both be considered, with the latter given enhanced consideration in younger patients. Screening through imaging may provide useful clues as to the correct preoperative diagnosis. The presence of fluid-fluid levels on a CT scan or MRI is a typical feature of ABCs, although it has also been reported in many other osseous lesions containing hemorrhage [22]. When fluid-fluid levels are demonstrated in FD, this outcome may be associated with simple cystic degeneration or sarcomatous transformation of FD [23].

In a literature review, the treatment of choice for an ABC with FD is total resection of the ABC and cranioplasty. Close follow up is mandatory due to the likelihood of recurrence of the lesion. Where possible, embolization of the feeding artery has been reported to reduce bleeding during surgery and enable maximal resection, as seen in our case [15].

We are reporting on a rapidly developing ABC in a pre-existing FD that presented with painful swelling and disfigurement in the right frontal region. Because subtotal resection is associated with a 50% recurrence rate, the patient underwent aggressive total resection of the ABC with partial removal of the FD followed by cranioplasty [24-27]. At the six-month postoperative follow up with CT and MRI images, there was no recurrence.

In conclusion, if a patient complains of a rapidly growing, soft mass with pain and pre-existing FD, secondary ABC development should be considered. In our case, preoperative angiography with embolization of the feeders was helpful in reducing intra-operative bleeding during an aggressive total resection.

Conflicts of Interest

The authors have no financial conflicts of interest.

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