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CASE REPORT

Unusual presentation of aneurysmal bone cyst (ABC) in children: pediatric intracranial osteosarcoma with secondary ABC

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Abstract

A 13-year-old female patient presented with painless vision loss and proptosis for 18 months. Imaging findings were highly suggestive of a supraorbital aneurysmal bone cyst (ABC) for which she underwent complete surgical excision. Postoperatively, she developed left hemiparesis. Computed tomography angiography (CTA) revealed right complete internal carotid arterial (ICA) thrombosis. This was managed conservatively, and she improved in hemiparesis over the next 3 weeks. Histopathology report revealed osteosarcoma with secondary ABC, for which she was referred for radiotherapy. At 1.5 months follow-up, the patient's left lower limb power improved to 4 + /5. She was walking without support, and her left upper limb power was 4/5.

Keywords Aneurysmal bone cyst · Osteosarcoma · Radiology · Proptosis · ICA thrombosis

Introduction

Aneurysmal bone cysts are generally benign lesions with multicystic appearance associated with extensive bony destruction. They can be of 2 types: primary and secondary. Primary and secondary ABCs are those without and with any other underlying benign or malignant pathology, respectively. Osteosarcoma is one of the causes of secondary ABC. Osteosarcomas in the pediatric population are most commonly located in the femur, followed by long bones and pelvis [1]; the intracranial site is very rare for primary osteosarcomas in children. Calcified brain metastasis without secondary ABC from primary osteosarcoma elsewhere has been reported [2, 3]. Primary osteosarcomas are rare intracranial tumors: 1–2% of all intracranial tumors [4]. We present an unusual presentation of pediatric osteosarcoma with secondary ABC in a 13-year-old girl.

Case presentation

A 13-year-old female patient was admitted with complaints of painless vision loss in the right eye with progressively increasing proptosis for 1.5 years. On examination, there was no restriction of eye movements in both eyes. Visual acuity in the right eye was a positive perception of light in all quadrants; left eye vision was normal. No neurocutaneous markers were present.

A plain X-ray skull (anterioposterior view) revealed a sclerosed sphenoid ridge and superior orbital wall (Fig. 1A). Computed tomography (CT) was suggestive of the destruction of the right superior orbital wall, anterior clinoid process, partial destruction of the inner table of the right frontal

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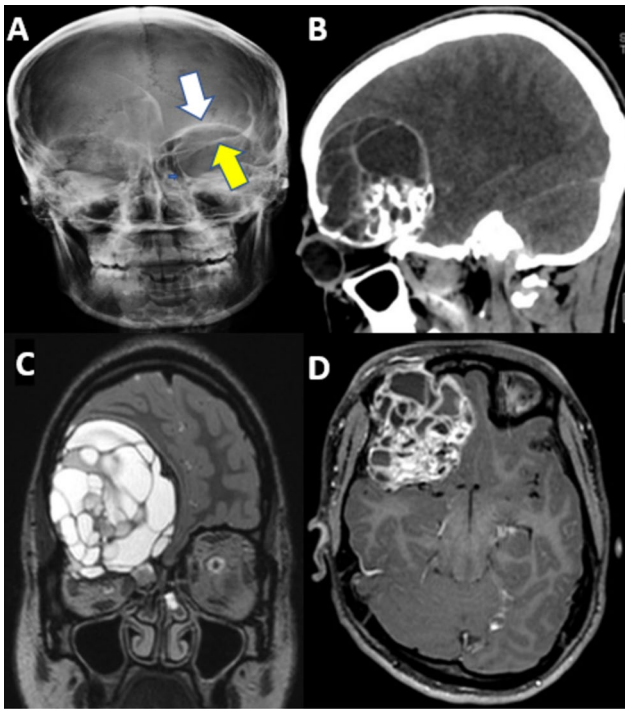

Fig. 1 Preoperative imaging. **A** X-ray skull AP view—sclerosed sphenoid ridge and superior orbital wall. A normal sphenoid ridge and an orbital wall on the left side are shown by yellow and white arrows, respectively. **B** CT lateral view—calcified lesion. **C** T2 MRI—multiseptated cystic lesion. **D** Contrast MRI—enhancement of septa

bone, and intralesional calcifications (Fig. 1B). Gadolinium-enhanced magnetic resonance imaging (MRI) of the brain and orbit revealed expansile, multilobulated, cystic septate lesion with internal fluid–fluid levels in right supraorbital region causing regional mass effect extending inferiorly into the orbit via superior orbital wall; inferomedially lesion was extending into the sphenoid sinus; and posteromedially abutting against anterior aspect of the cavernous sinus. There was the enhancement of septations and walls of the lesion on contrast images (Fig. 1C, D).

With a provisional radiological diagnosis of aneurysmal bone cyst (ABC), the patient underwent right fronto-orbital craniotomy and gross total excision of the lesion. The intraoperative impression of the operating team was that of ABC (Fig. 2A, B). However, the frozen section was suggestive of osteosarcoma; extensive resection with the drilling of surrounding partially involved bone and resection of calcified dura were done in view of the frozen biopsy report. The tumor was moderately vascular, septate, with multiple cysts containing altered blood. Venous ooze on the posteromedial aspect of the tumor cavity (cavernous sinus) was controlled using a thrombin-gelatin hemostatic matrix (Floseal® MATRIX).

The patient was extubated, fully conscious, and moving all four limbs. After 3–4 h postoperatively, she developed a left upper limb 0/5 and a left lower limb 3/5. Urgent head CT was done and revealed left motor cortex infarct; there

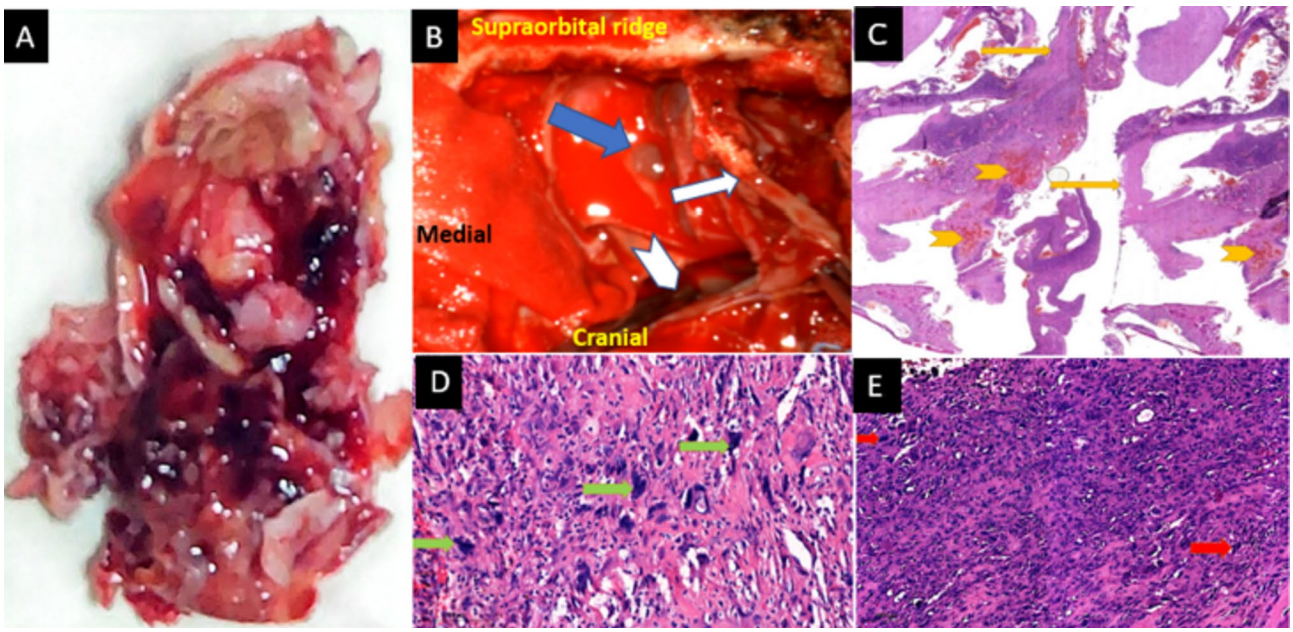

Fig. 2 **A** An intraoperative resected specimen of the calcified part of the tumor on which insertion of multiple septa is seen along with altered blood. **B** Intraoperative picture showing septa (white arrow), cystic cavity (white arrowhead), and intact cyst (blue arrow). **C** Low-power histopathology picture showing ABC-like areas with septa (yellow arrows) and hemorrhagic areas (yellow arrowheads). **D, E** High-power histopathology picture showing pleomorphic cells (green arrows) and atypical mitosis (red arrows) in the background of broad irregular trabeculae (osteoid)

was no operative site hematoma or mass effect (Fig. 3E). CT angiography was suggestive of abrupt cutoff of cervical ICA was seen just distal to bifurcation with non-visualization of the rest of cervical, petrous, laceral, and cavernous segment. Left common carotid artery (CCA) and its bifurcation, external carotid artery (ECA) and ICA, showed a normal course and contrast enhancement (Fig. 3A). Post-operative brain MRI (diffusion weight images) was suggestive of infarction at the right motor strip area (Fig. 3B, C) without any residual lesion. Still, persistent deformation of the right frontal lobe was noted even after gross total tumor resection (Fig. 3D). On post-operative day 3, the patient was started on antiplatelet and dyslipidemic medications. Her hemiparesis improved almost completely except for residual weakness in the left upper limb (power 4/5 MRC grade) on 3 months of follow-ups. Visual acuity status remained unchanged, and proptosis was resolved. The patient was referred for adjuvant radiotherapy after the final histopathology report (HPR) of osteosarcoma with secondary ABC. HPR was suggestive of blood-filled cystic spaces with fibrovascular septa. Septa were filled with pleomorphic cells with hyperchromatic nuclei with moderate to marked pleomorphism. Focally, osteoid was seen. Mitotic count was 3–4/10 high-power field. MIB-1 labeling index was approximately 18–20% in the highest proliferating area (Fig. 2C, D, E).

Discussion

Benign pathologies like osteoblastoma, chondroblastoma, giant cell tumor, fibrous dysplasia, and malignancies like osteosarcomas are known to be associated with secondary ABC. Giant cell tumor and fibrous dysplasia association are most common extracranially and cranially, respectively. Out of all ABCs, secondary and primary are 30% and 70%, respectively [5]. Few reports of intracranial osteosarcoma with secondary ABC are reported in adult patients [6, 7].

Eighty percent of primary ABCs occur before 20 years of age. Primary ABC generally has thinning of the cortical bone and expansion of lesions, which can be seen as soap bubble appearance on X-rays, and in T2-weighted MRI, they are seen as thin septa dividing multiple fluid-filled cystic cavities [7]. USP6 rearrangements in 65–70% and CDH11-USP6 fusion in 30% were seen recently in primary ABCs and not in secondary [5, 8]. Conversely, secondary ABCs are associated with particular tumor occurrence in terms of location and age known for that specific pathology. Aggressive tumors like osteosarcomas may have soft tissue mass and cystic cavities [7]. Our patients' imaging features looked more like primary ABC except for intralesional calcifications seen on CT (Fig. 1B). There was no soft tissue component either on MRI or during surgery to suspect it as a secondary ABC. Primary ABCs form from expansion and

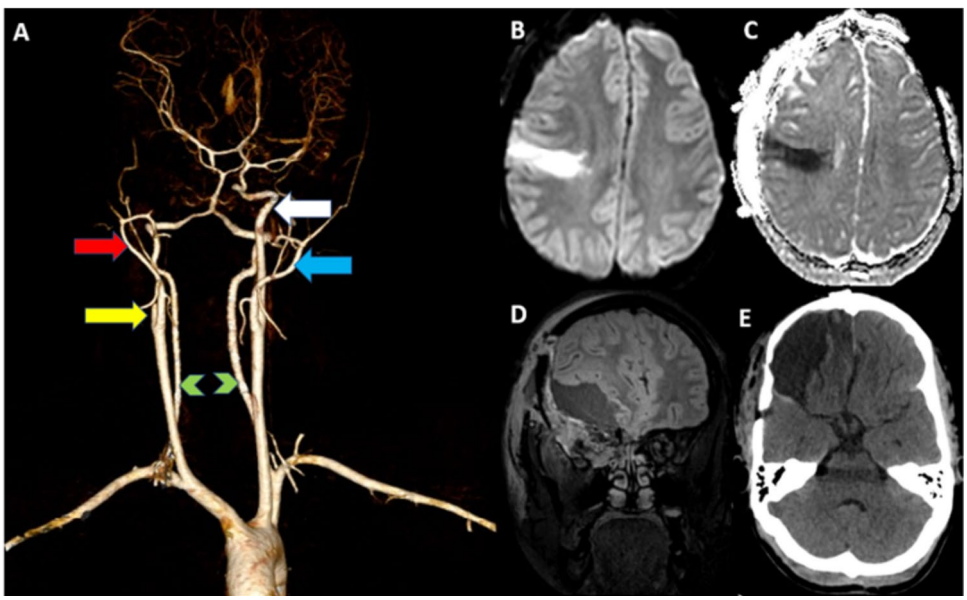

Fig. 3 Postoperative imaging. **A** CT angiography—Abrupt tapering of the right cervical ICA was seen just distal to the bifurcation (yellow arrow), right ECA normal course (red arrow), bilateral vertebral arteries' normal course (two green arrowheads), normal left ECA course (blue arrow), and normal left ICA course (white arrow). **B, C** Diffusion weight MRI image showing hyperintensity and corresponding apparent diffusion coefficient (ADC) image showing hypointensity suggestive of acute infarction at motor strip area. **D** Postoperative MRI coronal image showing the operative cavity without residual lesion. **E** Postoperative CT axial image showing operative cavity and persistence of deformed size of the right frontal lobe

Table 1 A literature review of pediatric secondary cranial ABCs

Sr. no	Author	Year	Pathology	Location in cranium	Age (years)	Sex	Size (cm)	Follow-up (f/u)
1	Gutierrez et al. [6]	2020	Chondromyxoid fibroma	Occipital bone	8	M	1×1×1	No f/u
2	Branch et al. [15]	1986	Fibrous dysplasia	Parietal bone	9	F	4×5	No f/u
3	Clavier et al. [16]	1988	Fibrous dysplasia	Temporal	2	M	6 (diameter)	No f/u
4	Wojno and McCarthy [17]	1994	Fibrous dysplasia	Temporal	14	F	5 (diameter)	No f/u
5	Haddad et al. [18]	1998	Fibrous dysplasia	Temporal	6.5	M	9×6	Four-year disease-free
6	Saito et al. [19]	1998	Fibrous dysplasia	Anterior skull base	2 cases (15 and 11)	Both F and M	-	3.5-year disease-free
7	Terkawi et al. [20]	2011	Fibrous dysplasia	Anterior skull base	7	F	-	Large recurrence after 5 months after surgery
8	Birk et al. [21]	2017	Fibrous dysplasia	Parietal	17	M	7×6×6	No f/u
9	Hadidy et al. [22]	2010	Fibrous dysplasia	Frontal	15	M	-	-
10	Manjila et al. [23]	2013	Fibrous dysplasia	Anterior skull base	10	M	3.8×3.6×2	Nine-month disease-free
11	Nasser [24]	2009	Psammomatoid ossifying fibroma	Frontal	12	M	4.5×4.1.5	No f/u
12	Roncaroli et al. [25]	2001	Eosinophilic Granuloma	Occipital	2	M	4 (Diameter)	14-month disease-free
13	Salmasi et al. [26]	2011	Fibrous dysplasia	Anterior skull base	16	M	-	Six-month disease-free
14	Sayama and MacDonald [27]	2010	Giant cell reparative granuloma	Petrous bone	16	F	3.8×2.8×0.5	32-month disease-free
15	Urgun et al. [28]	2016	Fibrous dysplasia	Occipital bone	14	F	-	No f/u
16	Cho et al. [29]	2017	Chondroblastoma	Sphenoid	13	M	4×4×3.5	One-year disease-free
17	Canzano et al. [30]	2021	Capillary venous malformation	Temporal	5	M	-	Two-year disease-free
18	Malik et al. [31]	2006	Fibrous dysplasia	Anterior cranial base	13	M	6×5.5	No f/u
19	Duarte et al. [32]	2019	Osteoblastoma	Occipital bone	4	M	7.5×5.8×5.2	One-month disease-free
20	Vargas [33]	2020	Cavernous hemangioma	Frontal	13	M	-	Three-month disease-free
21	Elleuch and Briki [34]	2016	Fibrous dysplasia	Parietal	16	F	6×4	Six-month disease-free
22	Park et al. [35]	2000	Langerhans cell histiocytosis	Parieto-occipital	7	M	7×7	-
23	Our case	2023	Osteosarcoma	Supraorbital	13	F	6×5×4.5	Six-week disease-free

bony destruction after increased intraosseous pressure due to regional vascular disturbances [5, 9]. The clinical course and histological features of primary ABC are better explained by the theory of dilated vascular spaces due to underlying arteriovenous anomaly [10]. The exact pathophysiology for transforming any malignant lesion into ABC-like changes is unknown. Telangiectatic osteosarcoma and ABC look similar radiologically and prognosis-wise, so they are generally misdiagnosed as ABCs [11].

Thrombin-gelatin hemostatic matrix is often used nowadays for attaining hemostasis in the cavernous sinus region during transcranial or endoscopic endonasal procedures

[12, 13]. We could not find the literature in which there was complete ipsilateral ICA thrombosis after thrombin-gelatin hemostatic matrix injection, causing ischemic events manifesting with severe neurological deficits postoperatively. Andrade-Barazarte et al. [14] reported complete ICA thrombosis after fibrin glue injection for cavernous sinus hemostasis during ICA aneurysm and petroclival meningioma surgery. According to them, probable mechanisms for this are (i) direct ICA injection, (ii) focal compression of cavernous ICA lumen, (iii) arteriovenous fistula between the cavernous sinus and ICA, and (iv) allergic idiosyncratic reaction to the glue. In our case, direct ICA injection is less likely as there was no brisk arterial bleeding near the cavernous sinus anytime during the surgery. We gave minimal compression at the venous oozing site for approximately 1 min using cottonoid, which is less likely to compress the ICA lumen to promote thrombosis. In our case, there may be the focal reaction of thrombin gelatin hemostatic matrix on the wall of cavernous ICA, causing focal thrombosis and ultimately causing retrograde thrombosis of the whole proximal ICA. During the literature review of pediatric cranial secondary ABCs, we found that the most common pathology was fibrous dysplasia, and the male-to-female ratio was 3.83:1 (Table 1).

Conclusion

Histopathology remains the gold standard in the diagnosis of primary ABC and in ruling out the possibility of secondary ABC in intracranial lesions. The frozen report is helpful in increasing the radicality of resection; the final HPE report helped in the timely referral of a patient for adjuvant radiotherapy. Internal carotid artery thrombosis secondary to the usage of thrombin gelatin matrix is very unusual; early diagnosis and treatment with antiplatelet agents helped in the resolution of thrombosis and uneventful recovery.

Author contributions Ninad Sawant wrote the manuscript, created the graphics, and reviewed the literature. Ninad Sawant and Harshavardhan Biradar contributed to the manuscript editing and literature review. Vivek Kumar created figures. Ninad Sawant made a table from the reviewed literature. Deepak Kumar Gupta, Ajay Garg, and Mehar Chand Sharma reviewed the manuscript, figures, and tables. Ajay Garg provided appropriate radiological images. Mehar Chand Sharma provided histopathological details. Deepak Kumar Gupta supervised.

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Declarations

Competing interests The authors declare no competing interests.

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