Solid variant of aneurysmal bone cyst on the cervical spine of a child: case report, differential diagnosis and treatment rationale

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Abstract

Introduction Despite numerous descriptive publications, the nature, character, differential diagnosis and optimal treatment of aneurysmal bone cysts (ABCs), remain obscure. The authors report a case of the solid variant of aneurysmal bone cyst (S-ABC) occurring in the posterior components and body of C7 vertebra focusing on the differential diagnosis and surgical treatment rationale.

Case report Right shoulder and neck pain were the presenting symptoms of 9-year-old boy. Torticollis developed

later on but no neurological deficit was found. Imaging revealed an osteolytic lesion with significant extraosseous extension. Although diagnosis favoured an ABC, imaging studies did not provide clear diagnostic criteria. CT guided biopsy performed preoperatively was also not directly diagnostic. Given that differential diagnoses included S-ABC but also giant cell tumor (GCT) of bone, decision was made to proceed with a staged, back and front, complete resection of the affected bony elements of C7. Preoperative spinal angiography showed supply to the tumor from the right ascending and deep cervical artery territories. Particle embolization was not performed due to the presence of ipsilateral supply to the anterior spinal artery at the C6 level and contralateral supply at the C7 level. Intraoperatively, histology taken from posterior elements, although again not clearly diagnostic, favoured S-ABC variant rather than GCT. Thus, initial plan was revised and anterior surgery was postponed as the extent of the dissection would have been dependent on the presumed diagnosis. The final histological report confirmed the diagnosis of an S-ABC. In view of this, it was decided to embolize the lesion to avoid a second stage anterior surgery. At embolization, repeat spinal angiography showed reduced tumor blush following the surgery. Distal branches of the deep cervical artery were occluded with platinum coils (avoiding the risks associated with the use of particles or liquid embolic agents). No further procedure was planned.

Conclusion Imaging and histological pattern of this specific type of ABC, differential diagnosis from GCT and the surgical protocol followed with the patient consist of an interesting case of revising the initial plan, according to the upcoming histological reports.

Keywords Solid variant of aneurysmal bone cyst · Cervical spine · Giant cell tumor · Differential diagnosis · Treatment rationale

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Case presentation

A 9-year-old boy was referred to our institution with an osteolytic lesion of C7 vertebra. He had right shoulder and neck pain almost 6 months prior to his referral. No specific history of injury or trauma was reported. As torticollis developed and the pain seemed to emanate from the neck rather than the shoulder, imaging was performed showing an osteolytic lesion of his cervical spine.

On his presentation to our outpatient department, he was in a soft collar. He denied any radicular symptoms and had no complaints of leg pain, weakness, numbness and balance dysfunction or bowel/bladder incontinence.

On physical examination he was tender in the lower cervical spine. Normal neurology was noticed in both, the upper and lower limbs with normal reflexes, no gait disturbances and signs of cord compression. His premedical history was unremarkable. Laboratory findings were all within normal limits.

Diagnostic imaging section

Historical review of the condition, epidemiology, diagnosis, pathology and differential diagnosis

ABC is an expansile, pseudotumoral lesion of unknown etiology, commonly occurring in the tibia, femur, pelvis, humerus and relatively rarely in the vertebral column representing approximately 15 % of all primary spine tumors [1–3]. The term was coined initially in 1942 by

Fig. 1 Preoperative anteroposterior X-ray

Fig. 2 Preoperative lateral X-ray

Fig. 3 Preoperative CT scan (sagittal view)

Jaffe and Lichtenstein [4] to describe a blood-filled lesion with a blow-out radiographic appearance. It is classified as *primary* if there is no underlying lesion or *secondary* when found in association with other tumors [2]. However, there is another distinct variant of ABC, the so-called solid variant, that was first described by Sanerkin et al. [5] in 1983.

Fig. 4 Preoperative CT scan (coronal view)

Fig. 6 Preoperative MRI scan (sagittal, T2-weighted image)

Fig. 5 Preoperative CT scan (axial view)

Fig. 7 Preoperative MRI scan (coronal, STIR image)

It is a rare lesion, accounting for 3.4–7.5 % of all ABCs, and only 15 cases (including our patient) occurring in the spine have been reported [6, 7]. Pediatric age group patients have been almost exclusively involved, ranging in age from 6 to 17 years (mean age 10.8 years) and the male:female ratio was 1:1.9. Pain (neck, back or chest) was the most common initial clinical symptom. Swelling, palpable tender mass, radiculopathy, myelopathy and scoliosis were less commonly reported signs/symptoms [7]. On average, symptoms persist for 12 months before definitive diagnosis [6].

Half of the cases occurred in the thoracic spine with the rest equally affecting the cervical and lumbar vertebrae. Like conventional ABCs, almost all S-ABC cases reviewed originated from the posterior elements of the vertebra. Involvement of the vertebral body was rare and reported in only three prior cases including our patient (Figs. 1, 2, 3, 4, 5, 6, 7, 8, 9, 10, 11). In all of these three cases the posterior elements were also affected.

Fig. 8 Preoperative MRI scan (axial, T1-weighted image)

Fig. 9 Preoperative MRI scan (axial, gadolinium-enhanced, T1-weighted image)

In terms of imaging, routine radiographic features on plain radiographs [6, 7] and CT scans [6] of the S-ABC include an osteolytic and expansile lesion that is indistinguishable from conventional ABC. Bone scintigraphy may reveal ring-like accumulation [8]—“donut sign” [9]—at the edge of the lesion. On the MRI scans, the S-ABC reveals homogeneous low-signal intensity on T1-weighted images and heterogeneous low-signal intensity with scattered high signal intensity areas on T2-weighted images with possible fluid–fluid levels [6]. This is similar to conventional ABC. However, conventional ABC’s septation seen in Gd-enhanced, T1-weighted images or T2-weighted images have never been reported in the solid variant [7]. Suzuki et al. [7] described homogenous high signal intensity throughout the lesion on Gd-enhanced MRI images,

Fig. 10 Angiography image (anterior spinal artery supply)

whereas in conventional ABC only the periphery of the lesion has that kind of signal.

On histological examination, a solid cystic variant is characterized by fibroblastic proliferation and absence of blood-filled spaces [1, 7]. The term S-ABC proposed by Sanerkin et al. [5] is histologically characterized by the following features: (1) florid fibroblastic or fibrohistiocytic proliferation without any cellular or nuclear pleomorphism, (2) areas rich in osteoclast-like giant cells, (3) osteoblastic differentiation with osteoid production, (4) aneurysmal sinusoids, and (5) occasional foci of degenerate calcifying fibromyxoid tissue. However, Suzuki et al. [7] in contrary described that osteoclast-like giant cells were seen only in minor foci, and the fibromyxoid tissue component was relatively abundant.

Depending on the proliferative component, the solid variant of ABC may be histologically mistaken for other benign tumors and tumor-like lesions, such as solitary bone cyst, hemangioma, chondroblastoma [6], osteoblastoma and giant cell tumor (GCT), which have to be ruled out histologically [7]. The lack or presence of anaplasia in histology studies argues against or support the diagnosis of a malignant tumor such as osteosarcoma or fibrosarcoma [7].

Rationale for treatment and evidence-based literature

Several reports on primary tumors of the spine in children have stated *the goals of treatment*. These should focus on

Fig. 11 Angiography image (deep cervical artery and anastomoses)

(1) obtainment of a definitive diagnosis through an appropriate biopsy or primary excision, (2) institution of appropriate surgical treatment as indicated by tumor type at the time of the initial procedure, (3) preservation or restoration of normal neurological function, and (4) maintenance of the growing spine stability to prevent subsequent secondary osseous and neurological dysfunction [10, 11].

The combination of plain X-rays, CT and MRI scans, usually support or is diagnostic in many cases. Nevertheless, any doubt about radiologic diagnosis makes *biopsy* mandatory [2]. Accurate histological evaluation with correlation of radiographic and MRI findings is imperative for definitive diagnosis [12]. In lesions with a major hemorrhagic component, needle biopsy can be misleading; therefore, an open biopsy is suggested. In these cases, appropriate surgical treatment should be performed at the time the biopsy is done [2].

Despite the lack of clear treatment guidelines about S-ABC, adequate surgery appears to be the mainstay of treatment reported in the literature, especially in cases of neurological compromise (radiculopathy or myelopathy) [6, 7]. *Curettage* is often the treatment of choice for conventional ABCs [1–3, 13, 14] but also for S-ABCs [6, 7] and describes the piecemeal removal of the tumor. As such, it is always an intralesional procedure [15]. It is a commonly performed procedure because of the benign character of the lesion, although recurrence may develop [8].

En bloc surgery although not reported in the literature for the treatment of S-ABCs provides the surgeon with

Fig. 12 Pre-embolization image (deep cervical artery)

another option and has the lowest rate of local recurrence for ABC patients. Considering the hemorrhagic nature of the tumor, it is indicated for posteriorly located conventional ABCs—to avoid excessive bleeding that may accompany intralesional excision—if embolization could not be performed or after local recurrence [2, 3, 10]. Bone grafting was also used for promoting fusion in the resection bed and/or restoring stability of the spine.

Embolization of feeding segmental arteries has been proposed as a preoperative adjunct to surgery in order to reduce blood loss or decrease contamination potential of tumor for primary or secondary ABCs. As primary treatment could be the procedure of choice for conventional ABCs in specific cases excluding patients with high risk of pathologic fracture, neurological involvement or tumors located at the cervicothoracic spine because of the risks of cord ischemia and ectopic embolization [2]. Despite these concerns, the literature [6] suggests angiography and embolization can be performed without a significant risk of permanent neurological deficit, skin, or muscle necrosis. Until now there is no evidence in the literature about embolization use in treating patients with S-ABCs.

Radiation therapy has been reported in the literature as an adjuvant treatment type of S-ABCs [6]; reports of late post-irradiation sarcomas, myelopathy and deformity [3, 7] in patients with conventional ABC have made other authors more cautious about its use especially in children [2], and radiation therapy should be reserved for patients with inoperable lesions because of location or associated medical conditions, or aggressive recurrent disease.

In our patient, plain X-rays and scans were supportive but not diagnostic of ABC. A CT guided biopsy was performed

Fig. 13 Post-coiling image

Fig. 14 Post-coiling image (anterior spinal artery filling)

and two samples were taken. Histological examination found the biopsy material to consist predominantly of blood clot and few cellular fragments consisting of multinucleated osteoclast type giant cells intermingled with spindle shaped

Fig. 15 Two-year follow-up MRI scan (sagittal, T2-weighted image)

mononuclear stromal cells. The features were in accordance with a benign giant cell rich lesion and the differential diagnoses included S-ABC and GCT of bone.

Differential diagnosis was S-ABC/GCT. Although S-ABC of the spine mainly affects pediatric age group patients (mean age 10.8 years) with male:female ratio of about 1:1.9, GCT of the spine occurs most commonly between the ages of 11–61 years of age (average 30 years) [16]. However, affected male:female ratio is the same as S-ABCs [17]. Patients with GCT of the mobile spine seem to present later in the course of the disease than those with S-ABC with a higher incidence of neurological signs and symptoms, approaching up to 46 % of cases [17]. Typically S-ABCs originate in the posterior neural arch and expand unilaterally to produce an eccentric paravertebral lesion [7]. Vertebral body involvement is rare and as a result body collapse not often seen on X-rays. On the other hand, GCTs usually arise from the vertebral body and spare the posterior elements [16, 18]. Half of the S-ABC cases reviewed in the literature occurred in the thoracic spine (57 % of the cases) with the rest equally affecting the cervical and lumbar vertebrae. In 2006, Turcotte et al. [18] reported on 13 cases of spinal GCT. Six of them affected the lumbar spine, four the cervical, two the thoracic while in one case the cervicothoracic junction was involved. Boriani et al. [16], however, reviewing 49 GCTs of the mobile spine stated that the thoracic and lumbar spine was involved almost equally (44.9 and 46.9 %, respectively) while the cervical spine was involved in 12.25 % of the patients.

Fig. 16 Two-year follow-up MRI scan (coronal, STIR image)

Radiographic imaging hallmarks of GCTs include osteolysis and expansion without a marked sclerotic border. Bone scintigraphy reveals homogeneous uptake or a nonspecific “donut sign” as a rim of hyperintensity surrounding central osteopenia [9]. CT shows the lack of mineralizing matrix and may reveal a thin sclerotic border surrounding the soft tissue component, reflecting intact periosteum. Secondary aneurysmal bone may form due to hemorrhage and may be evident on CT or MRI as fluid–fluid levels [9]. MRI reveals intermediate T1-weighted signal with hyperintense hemorrhagic regions and hypointense cystic regions while on S-ABCs homogeneous low-signal intensity is seen [6]. Solid regions are often hypointense on T2-weighted images, reflecting a high collagen content [9].

Microscopic features of GCT include oval-to-spindle shaped mononuclear stromal cells and also variable multinucleate osteoclast-like giant cells. Reactive fibrohistiocytic proliferation is common [9]. S-ABC has similar histological characteristics but further more osteoblastic differentiation with osteoid production, aneurysmal sinuoids, and occasional foci of degenerate calcifying fibromyxoid tissue co-existing. Distinguishing features of S-ABC and GCT are not clear in the literature and rely on the experience of the pathologist.

Given that CT guided biopsy was not clearly diagnostic we decided to aggressively treat the patient as suffering from GCT to minimize recurrence rates as local recurrences of the spine are much more difficult to treat than of

Fig. 17 Two-year follow-up MRI scan (axial, T2-weighted image)

Fig. 18 Two-year follow-up MRI scan (axial, T1-weighted image)

the extremities [19]. We planned a staged back and front intralesional en bloc excision. We decided to perform the posterior procedure initially to resect the bony elements of C7, obtain new histology, and stabilize the spine. Anterior procedure would have been mandatory, if GCT was confirmed, to excise the remaining part of the tumor.

To reduce intraoperative bleeding, angiography was performed preoperatively aiming to embolize the tumor. Unfortunately, angiography revealed right-sided tumor supply from the deep cervical and ascending cervical territories and left C7 and right C6 radiculomedullary supply thus contra-indicating particle embolization (Figs. 10, 11).

Fig. 19 Two-year follow-up MRI scan (axial, gadolinium-enhanced, T1-weighted image)

Fig. 20 Two-year follow-up anteroposterior X-ray

Procedure (surgery, intervention)

After explaining in detail various treatment alternatives to the patient and his parents, we performed surgical excision of the bony lesion. This process consisted of intralesional en bloc excision [15] of the lesion within the lamina and spinous process of C7. Through a posterior mid-line skin incision, the C7 lamina was removed through a left hemilaminectomy and subsequent removal of the right hemilamina in one piece exposing the lesion in the lateral mass. Right vertebral artery was occluded by the tumor. Both C7

Fig. 21 Two-year follow-up lateral X-ray

and C8 nerve roots were explored. The inferior aspect of the C6 lamina was also resected as well, removing all microscopic evidences of tumor. Extension of the tumor into the pedicle to the C7 vertebral body was noticed. The lateral aspect of the C7 vertebral body was exposed. Diluted peroxide irrigation was performed. There was no microscopic tumor visible with the exception of the soft tissue anterior to the plane of the nerve roots. Instrumented posterior cervical fusion C6–T1 was performed. Pedicle screws were inserted on both sides of T1 and on the right side of C6 vertebrae while lateral mass fixation on the left side of C6 was done. That was decided in order not to risk endangering the remaining left vertebral artery. A rib strut removed subperiosteally from T10, was split in two parts and fixed over the C6 and T1 laminae to promote fusion. The intraoperative histology had shown isolated giant cells. In discussion with the lab during the time of surgery, it could not be verified that this was in accordance with a GCT or an S-ABC, whereby the latter was favoured but further opinion would be needed. The plan therefore was to await confirmation of histology prior to the anterior surgery as the extent of the dissection will be dependent on the presumed diagnosis.

The final histology reports favoured a S-ABC over a GCT. As such we tried a repeat angiography aiming to embolize the residuum of the tumor. The idea was that the embolization will suffice in preventing further growth of the aneurismal bone cyst thus avoiding a second stage procedure. Branches of the right deep cervical artery were

explored with an echelon microcatheter and small pedicles supplying dysplastic tumor circulation were occluded with hydrosoft coils (instead of using particles or glue in view of the potential anastomosis with the anterior spinal and vertebro-basilar circulations). Subsequent runs showed a reduction in flow within the dysplastic tumor circulation (Figs. 12, 13, 14).

Procedure imaging section

Outcome, follow-up

Patient at the 2 year follow-up appointment was clinically very well with his neck straight, and no complain of any pain or any neurological symptoms. Plain X-rays show a degree of calcification of the previous cyst and fusion in alignment on the lateral view. On his MRI scan, the component within the expanded right C7 transverse process has reduced in size and has imaging features consistent with the plain film findings (progressive ossification/matrix formation). However, the small cystic component within the lateral aspect of the C7 vertebral body has increased in size. There was an adjacent flow void suggesting this is likely to represent a small residual component of ABC. Suzuki et al. [7] in 2004 reported on spontaneous regression of not fully extirpated S-ABCs. In keeping with that, the further plan was to follow the patient after a 6-month period, performing a follow-up MRI scan prior to his next consultation while he was advised to return back to full activities (Figs. 12, 13, 14, 15, 16, 17, 18, 19, 20, 21).

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References

1. Mankin HJ et al (2005) Aneurysmal bone cyst: a review of 150 patients. *J Clin Oncol* 23(27):6756–6762

2. Boriani S et al (2001) Aneurysmal bone cyst of the mobile spine: report on 41 cases. *Spine (Phila Pa 1976)* 26(1):27–35
3. Saccomanni B (2008) Aneurysmal bone cyst of spine: a review of literature. *Arch Orthop Trauma Surg* 128(10):1145–1147
4. Jaffe JL, Lichtenstein L (1942) Solitary unicameral bone cyst with emphasis on the roentgen picture. *Arch Surg* 44:1004–1025
5. Sanerkin NG, Mott MG, Roylance J (1983) An unusual intraosseous lesion with fibroblastic, osteoclastic, osteoblastic, aneurysmal and fibromyxoid elements. “Solid” variant of aneurysmal bone cyst. *Cancer* 51(12):2278–2286
6. Al-Shamy G et al (2011) Solid variant of aneurysmal bone cyst of the thoracic spine: a case report. *J Med Case Reports* 5(1):261
7. Suzuki M et al (2004) Solid variant of aneurysmal bone cyst of the cervical spine. *Spine (Phila Pa 1976)* 29(17):E376–E381
8. Sato K et al (1996) Solid variant of an aneurysmal bone cyst (giant cell reparative granuloma) of the 3rd lumbar vertebra. *Nagoya J Med Sci* 59(3–4):159–165
9. Dickman CA, Fehlings M, Gokaslan ZL (2006) Spinal cord and spinal column tumors: principles and practice. Thieme
10. Khalil IM et al (2006) Aneurysmal bone cyst of the cervical spine in a child: case report and review of the surgical role. *Surg Neurol* 65(3):298–303 (discussion 303)
11. Weinstein JN, McLain RF (1987) Primary tumors of the spine. *Spine (Phila Pa 1976)* 12(9):843–851
12. Cottalorda J, Bourelle S (2006) Current treatments of primary aneurysmal bone cysts. *J Pediatr Orthop B* 15(3):155–167
13. Beiner JM et al (2006) An aneurysmal bone cyst in the cervical spine of a 10-year-old girl: a case report. *Spine (Phila Pa 1976)* 31(14):E475–E479
14. Ramme AJ, Smucker JD (2011) Balancing spinal stability and future mobility in the cervical spine: surgical treatment of a case of osteoblastoma with secondary aneurysmal bone cyst. *Spine J* 11(5):e5–e12
15. Boriani S, Weinstein JN, Biagini R (1997) Primary bone tumors of the spine. Terminology and surgical staging. *Spine (Phila Pa 1976)* 22(9):1036–1044
16. Boriani S et al (2012) Giant cell tumor of the mobile spine: a review of 49 cases. *Spine (Phila Pa 1976)* 37(1):E37–E45
17. Martin C, McCarthy EF (2010) Giant cell tumor of the sacrum and spine: series of 23 cases and a review of the literature. *Iowa Orthop J* 30:69–75
18. Turcotte RE (2006) Giant cell tumor of bone. *Orthop Clin North Am* 37(1):35–51
19. Balke M et al (2012) Giant cell tumors of the axial skeleton. *Sarcoma* 2012:410973