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Fibromyxoma of the Petrous Bone: A Case Report

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Abstract:

Myxomas are uncommon tumors typically located in the left atrium of the heart, bones of the jaw or skeletal muscle. We present a rare case of fibromyxoma in the petrous bone. A 9-year-old boy complained of a protrusion and tenderness in the left postauricular area. Neurological and otorhinolaryngological examination did not reveal any abnormality. Computed tomography and magnetic resonance imaging revealed a well-demarcated, solid tumor with a size of 62×92×80 mm. Calcification and petrous bone destruction was evident, and the lesion showed heterogeneous enhancement with contrast media. Thallium-201 single photon emission computed tomography (SPECT) and Technetium-99m muramyl dipeptide bone scintigraphy revealed high uptake in the tumor. Early and delayed images of technetium-99m-diethylene triaminepentaacetic acid-human serum albumin SPECT disclosed no definite uptake in the tumor. Cerebral angiography revealed marked extracerebral mass effect and mild vascular supply of the tumor from the left ascending pharyngeal artery, suggesting occlusion of the left sigmoid sinus. Total radical resection of the tumor was carried out. The tumor was totally extradural, yellow-whitish in color, elastic soft, jelly-like, bloodless and resectable. The histological diagnosis was fibromyxoma.

Key words: fibromyxoma, petrous bone, bone tumor, computed tomography, magnetic resonance imaging, single photon emission computed tomography

Introduction

Myxomas are uncommon tumors, generally documented as being located in the left atrium of the heart, bones of the jaw or skeletal muscle[13,23,25]. Myxoma or fibromyxoma of the bones in the head and neck usually appear in the maxilla and/or mandible. Fibromyxoma of extragnathic bones is a very rare and controversial entity[2]. Due to that rarity, to date, only one case of fibromyxoma of the petrous bone has been previously reported in the literature[22]. We here report a second case.

Case Report

A 9-year-old boy complained of a protrusion and tenderness in the left postauricular area. The size of the protrusion was approximately 2×2 cm. Neurological examination showed no deficits. Ocular fundus examination revealed bilateral papilledema. An audiogram demonstrated normal hearing, and otorhinolaryngological examination did not reveal any abnormality. A skull radiography revealed marked erosion of the petrous bone on the left side (Fig. 1).

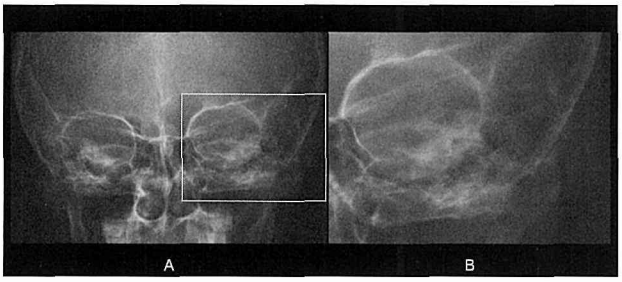

Fig. 1 (A) A skull X-ray showing destruction of the petrous bone on the left side.
(B) Magnification of the left petrous bone.

Computed tomography (CT) revealed a low density, well-demarcated solid lesion, with a dimensional size of 62×92×80 mm. Calcification and petrous bone destruction was evident, and the lesion showed relative enhancement with contrast media except at the center of the tumor (Fig. 2). Erosion of the temporal bone did not extend to the semicircular canals or the middle ear (Fig. 3).

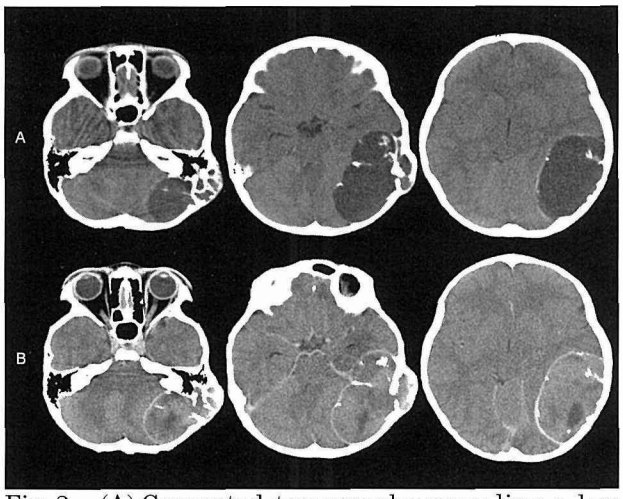

Fig. 2 (A) Computed tomography revealing a low density well-demarcated solid tumor with calcification and destruction of the left petrous bone.
(B) The tumor enhanced with contrast media (except for the center).

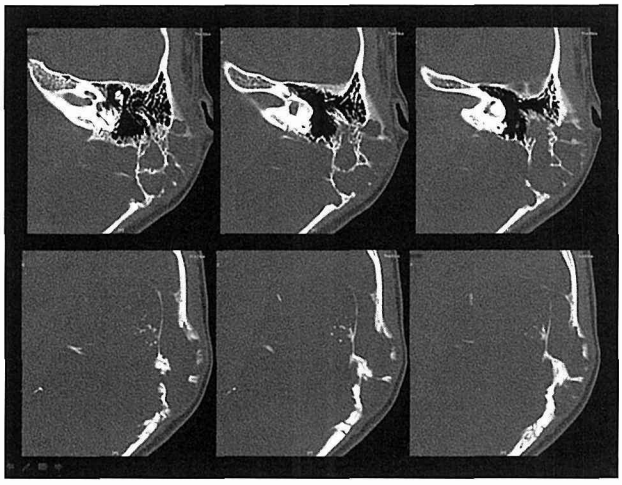

Fig. 3 A bone window CT scan showing the remaining semicircular canals.

Magnetic resonance (MR) imaging revealed the tumor as hypointense in T1 and FLAIR images, extremely hyperintense in T2 images and isointense in proton density and diffusion weighted images (Fig. 4). Gadolinium (Gd)-enhanced MR imaging showed heterogeneous enhancement of the tumor (Fig. 5). Thallium (Tl) 201 single photon emission computed tomography (SPECT) revealed high uptake in the tumor (Fig. 6). Conversely, both early and delayed

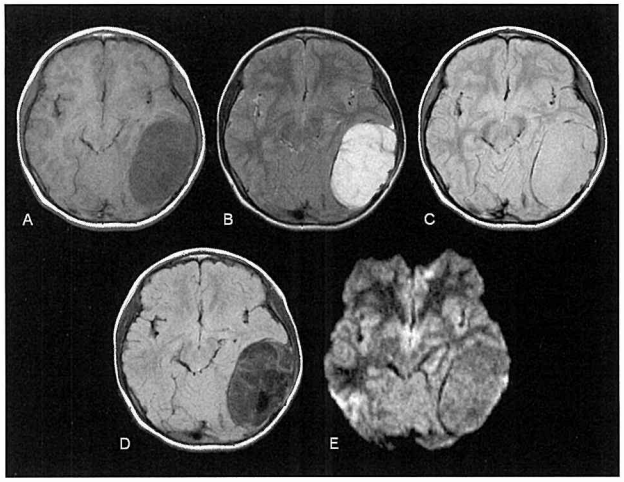

Fig. 4 Magnetic resonance imaging disclosing the tumor as hypointense on the T1 weighted image (A), extreme hyperintense on the T2 weighted image (B), isointense on the Proton dense image (C), hypointense on the FLAIR image (D), isointense on the diffusion weighted image (E).

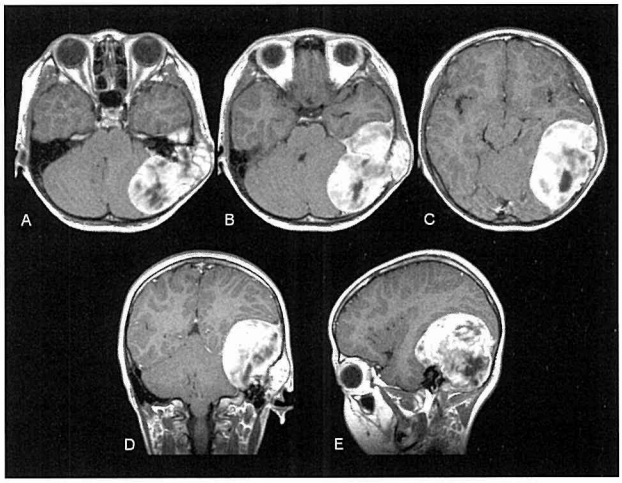

Fig. 5 Gadolinium-enhanced T1 weighted MR imaging showing heterogeneous enhancement of the tumor. (A-C) axial image, (D) coronal image, (E) sagittal image.

images of technetium-99m-diethylenetriaminepentaacetic acid-human serum albumin (Tc-99m HSAD) SPECT disclosed no definite uptake in the tumor (Fig. 7). Technetium-99m muramyl dipeptide (Tc-99m MDP) bone scintigraphy showed a hot area in the left temporal bone (Fig. 8). Cerebral angiography revealed marked extracerebral mass effect and mild vascular supply of the tumor from the left ascending

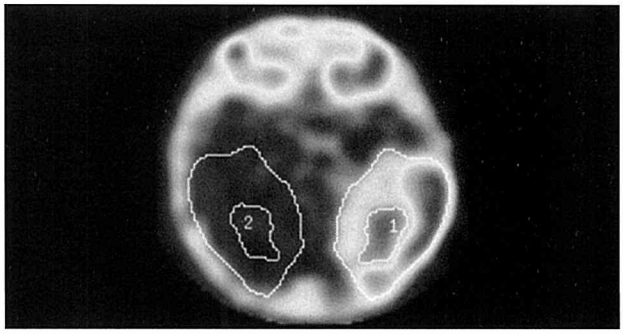

Fig. 6 Thallium (Tl)-201 single photon emission computed tomography (SPECT) revealing high uptake in the tumor. The ratios of Tl-201 uptake in the tumor to uptake in the corresponding normal contralateral areas (uptake index) was 2.3.

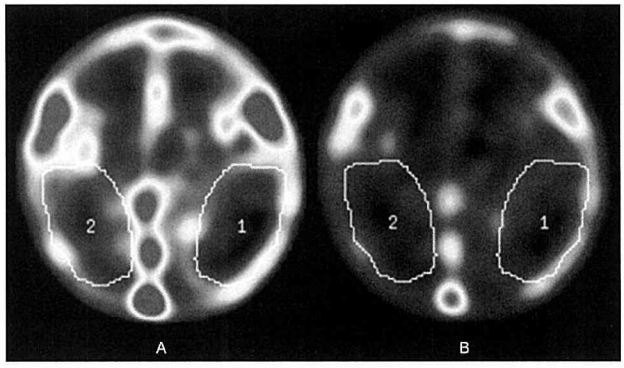

Fig. 7 Technetium-99m-diethylene triaminepentaacetic acid-human serum albumin (Tc-99m HSAD) SPECT disclose no definite uptake into the tumor in the (A) early and (B) delayed images.

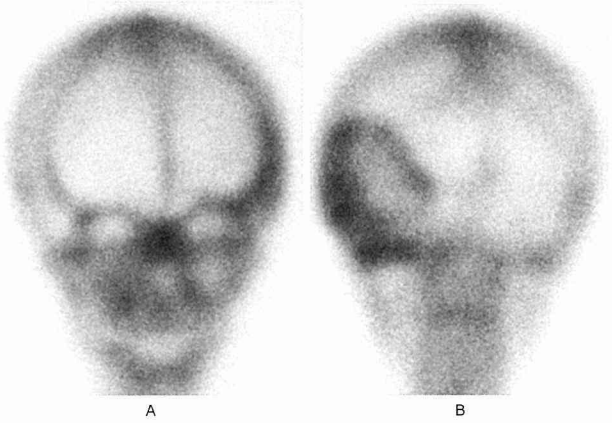

Fig. 8 Technetium-99m muramyl dipeptide (Tc-99m MDP) bone scintigraphy showing hot area in the left temporal bone.
(A) frontal view, (B) posterior view

pharyngeal artery, suggesting occlusion of the left sigmoid sinus (Fig. 9).

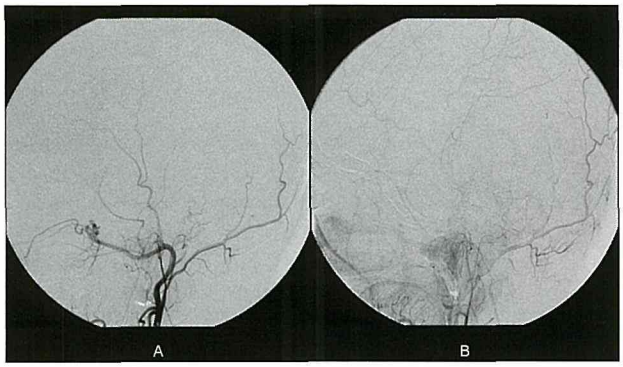

Fig. 9 Left external carotid arteriography demonstrating mild vascular supply of the tumor from the left ascending pharyngeal artery and posterior auricular artery, suggesting occlusion of the left sigmoid sinus.
(A) arterial phase, (B) venous phase

Total radical resection of the tumor was carried out. The tumor invading the mastoid air sinus was totally extradural, yellow-whitish in color, elastic soft, jelly-like, bloodless and resectable (Fig. 10). Postoperative CT and MR imaging showed that the tumor was totally resected (Fig. 11).

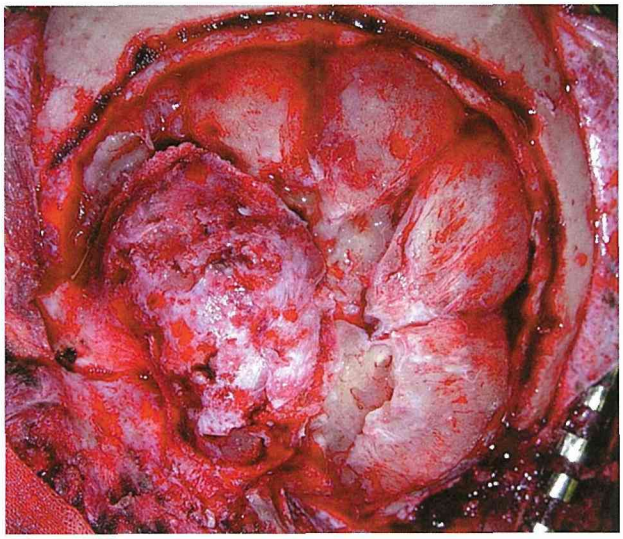

Fig. 10 An intraoperative photo after craniotomy showing extradural, yellow-whitish, elastic, soft tumor in clump-like configurations.

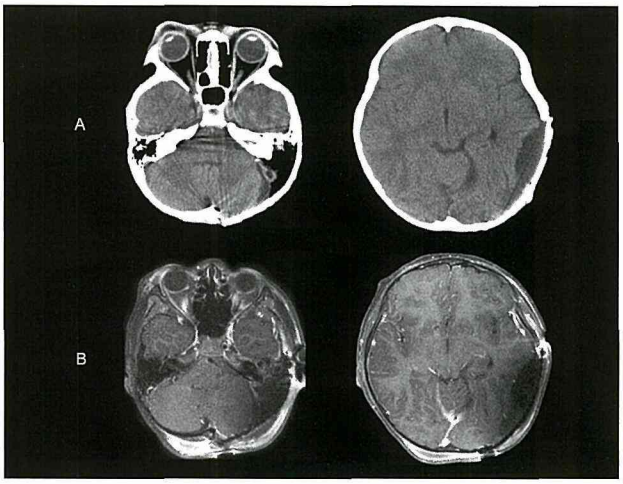

Fig. 11 Postoperative CT (A) and MR (B) images showing that tumor has been totally resected.

Microscopic examination with hematoxylin and eosin (H&E) stains revealed that the tumor was comprised of short spindle cells in a loose, myxoid background with foam cells (Fig. 12). The myxoid tissue contained a number of partially fibrous tissue bands. Minimal bony tissue existed in the tumor. Similarly, no chondroid tissue or nerve tissue was noted in the

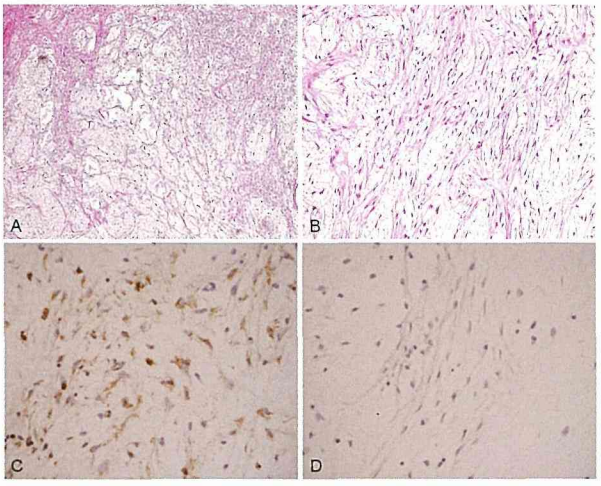

Fig. 12 Photomicrographs showing short spindle cells with loose, myxoid tissue and partially fibrous tissue bands with minimal bony tissue, and no chondroid or nerve tissue in the tumor.
(A) Hematoxylin and eosin (HE) stain (×40),
(B) HE stain ×200), (C) Immunohistochemical staining was positive for vimentin, (D) negative for S-100 protein

tumor. Immunohistochemical stainings revealed the tumor cells were positive only with vimentin and negative with S-100 protein, PAS, Alcian blue, α-SMA and CD68. Staining with CD68 showed positive only in the macrophages. A histological diagnosis of fibromyxoma was thus able to be made.

Discussion

Historically, myxomas have been reported and discussed for almost 100 years. Myxoma of the bone was initially reported by Bloodgood[5], but the tumor in the reported case had potential for metastasis and was likely a chondromyxoid fibroma, or chondrosarcoma with myxoid features. Bullough and Jaffe's definition of the pathological criteria for myxoma of the appendicular skeleton was reported by Stout in 1948 and Bauer in 1954[4,14,16,23]. A case of myxoma of the toe bone described by Perou, et al. (1967) is recognized as the first, true, documented myxoma[2,18,19].

Fibromyxomas have a similar, if not shorter, history. A case of fibromyxoma of the tibia was initially reported by Lehmann, and three other cases were reported by Marcove, et al. who defined the pathological criteria of fibromyxoma of bone in their article in 1964[15,16]. Some other earlier case reports on fibromyxomas were more recently re-diagnosed as other tumor entities, such as chondromyxoid fibromas, ganglionic cysts, or ganglia[4,8,10,20,21]. Chronologically, reports of fibromyxomas of the extragnathic bones appeared next. Two cases in the femur were reported by Caballes, one by Adler, another one by Goldman, and two cases in the pelvis and cervical vertebra were discussed by Abdelwahab[1,2,7,11].

Myxomas or fibromyxomas are also some of the most common tumors of the jaw, and account for between 50 to 70% of all mandibular tumors[3]. Such mandibular lesions usually occur in the second decade of life[17]. Extragnathic myxomas or fibromyxomas, on the other hand, are very rare.

In the case of the temporal bone, two myxomas were reported by Bulghov and Charabi, but to date, there has only been one instance reported in the literature concerning the occurrence of a fibromyxoma in the temporal bone[6,9,22]. The first was reported by Srinivasan, and we present the second case here.

Various imaging modalities help diagnose and delineate tumor types. In epidermoid tumors, diffusion weighted MR images are hyperintense and do not need to be gadolinium-enhanced. Meningiomas yield a less hyperintense T2 image. In condromas and chondrosarcomas, the CT reveals the tumor as isodense to high dense, and chordomas appear at a more midline location in the skull. In giant cell tumors, T1 MR images are isointense, and T2 images are hypointense. In eosinophiric granulomas, T1 images are isointense to hyperintense. A CT scan will reveal an osteoma as high dense or isodense, and cholesterol granulomas show a hyperintense T1 image. Ewing's sarcoma is characterized by isodense to high dense areas in a CT scan, and homogeneously enhanced by contrast media.

In the case reported by Srinivasan, the tumor revealed expansive osteolysis at the petrous apex by CT scanning, with MR imaging yielding hypointense T1 images and extremely hyperintense T2 images[22]. In our case, MR imaging disclosed the same intensity in T1 and T2 images. In addition, we also used and reported other sequences of MR images, SPECT, bone scintigrams and angiograms in the work-up for this tumor.

Fibromyxomas consist of fibrous and myxoid stroma with a varying degree of calcification and/or ossification. Fibromyxomas do not exhibit a lobulated pattern of chondromatous elements, and a negative staining of S-100 can be used to verify their difference to chondromas and chondrosarcomas.

The local recurrence rate of bone myxoma is approximately 25% after curettage surgery[12,24]. Furthermore, radiation therapy has not been found to be effective for this tumor. The recurrence rate after complete resection must reasonably be lower than that after just curettage surgery, strongly making the case that this should be the preferred treatment

modality in such benign tumors.

Conclusion

We reported a case of fibromyxoma of the petrous bone. This is the second such case of temporal fibromyxoma.

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