Protuberant Fibro-osseous Lesion of the Temporal Bone: “Bullough Lesion”

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Abstract: Primary tumors of the calvarium are infrequent, and with the exception of osteoma, lesions confined to the surface of the skull are very rare. The differential diagnosis includes benign and malignant matrix forming tumors, other mesenchymal tumors, and reactive lesions. Fibro-osseous lesions are characteristically centered within bone and surface fibro-osseous lesions always prompt consideration of parosteal osteosarcoma, which is rare but well documented in the calvarium. We present 2 cases of a distinctive lesion of the temporal bone intimately related to the occipito-mastoid suture and typically presenting as a retroauricular soft tissue mass with calcific densities, confined to the soft tissues on the outer table of the skull without intraosseous involvement. The lesion is characterized histologically by rounded and ovoid zones of ossification within a bland fibrous stroma. The first 2 cases were documented in 1999 as “Protuberant fibro-osseous lesion of the temporal bone.” We present a further 2 cases, 1 of 2 years duration and the other with a 10-year history. This distinctive entity, which must be distinguished from other fibro-osseous lesions, including subtle low-grade parosteal osteosarcoma, seems to behave in a benign fashion and thus far recurrence is not documented. Local excision seems adequate. The pathologic features in the original report were documented by Prof Peter Bullough. As these cases were recognized by him alone we propose calling this entity “Bullough lesion” or, better still, “Bullough’s Bump!”

Key Words: temporal bone, mastoid, fibro-osseous, protuberant, tumor

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Common matrix producing lesions of the skull and craniofacial bones include osteoma, fibrous dysplasia, and less frequently, osteoblastoma. In the mandible and maxilla, lesions related to the dental apparatus including ossifying fibroma, forms of cemental dysplasias, and cementoblastoma must also be considered. Fibro-osseous lesions of the skull and craniofacial bones most often involve the mandible and maxilla, and less commonly affect nasal bones, sinuses, occiput, and temporal bone.

Clinical, pathologic, and radiologic correlation is required for a definitive diagnosis, and occasionally these lesions may simulate a primary malignant bone tumor, particularly in the case of osteoblastoma and ossifying fibroma.

We present 2 cases of a very rare but distinctive lesion originally documented in 1999 and designated “protuberant fibro-osseous lesion of the temporal bone.”

These are benign, slow-growing lesions composed of a bland fibrous stroma in which rounded or ovoid zones of ossification are present. They are confined to the surface of the temporal bone overlaying the mastoid air cells, and seem to represent a unique entity clinically, radiographically, and pathologically. The combined radiographic and histologic changes may prompt consideration of a variety of lesions including low-grade parosteal osteosarcoma, from which this lesion must be distinguished.

CASE REPORTS

Case 1

A 27-year-old man presented with a localized right retroauricular swelling tender to pressure noted for 1 year. There was no antecedent trauma, or history of radiation exposure. The swelling was 2 cm in diameter, firm to hard in consistency, and fixed to the underlying skull. The overlying skin was stretched, smooth, and nonadherent.

There was no cervical lymphadenopathy. General physical examination including neurologic examination was unremarkable. Hematologic and biochemical parameters were normal.

Roentgenograms of the skull showed a mass in the right mastoid area in which widespread densities were present giving a speckled appearance (Fig. 1A).

CT of the skull revealed a right retro-auricular mass with some mineralized matrix production lying in the soft tissues overlaying the mastoid outer table of the skull closely related to the occipito-mastoid suture (Fig. 1B). The mass was predominantly low density and expanded with calcified foci centrally and a faint incomplete thin calcified rim. The central mineralized foci were largely rounded. The cortex on which the lesion lay was

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FIGURE 1. A, Plain x-ray in case 1 reveals the mass containing rounded mineralized densities within the soft tissues overlying the mastoid air cells (arrow). B, Axial CT scan in Case 1 shows a soft tissue mass posterior to the mastoid process with fine rounded densities and a subtle partially calcified peripheral rim. C, At low power on the left of the image variably sized ovoid osseous structures lie within a fibrous stroma, the larger one at the top left composed predominantly of lamellar bone. The stroma is in direct continuity with the outer aspect of the cortex, which is of slightly irregular contour, the collagen of the fibro-osseous lesion merging with the outer aspect of the cortex through a calcified tide mark. A mastoid air cell is included at the bottom right. D, This rounded zone of ossification is composed of moderately cellular immature woven and coarse lamellar bone within bland sparsely cellular fibrous stroma. Osteocyte nuclei are inconspicuous and osteoblastic activity is absent. E, Polarisation microscopy confirms the presence of a mixture of mature lamellar, coarse lamellar, and focal woven bone. In some osseous structures osteoblastic activity is noted and an occasional resorption pit and irregular cement lines denote osteoclastic activity. F, In the most cellular areas of stroma the fibroblastic cells have a somewhat fasciculated arrangement. Insert: Stromal fibroblastic cells have bland nuclei with regular nuclear contours and small inconspicuous nucleoli. Mitotic activity is not seen. Scattered mast cells are present.
intact but more irregular in contour than the adjacent cortex. There was no apparent intramedullary or intracranial extension. The general impression was of a nonaggressive lesion and although the appearances were considered unusual, the differential diagnosis included a variety of lesions including osteoblastoma, juxta cortical chondroma, myositis ossificans, or an unusual ossifying haemangioma. A parosteal osteosarcoma was considered less likely.

Local excision was carried out. A 2-cm diameter lesion of firm to hard consistency was located on the outer table of the skull and was easily excised with the aid of an osteotome and a drill. There was no evidence of inner table breach.

**Pathologic Examination**

The gross specimen comprised a fragment of bony tissue 17 × 15 × 5 mm. Microscopically the tissue was composed of a strip of mature cortical bone, the inner aspect of which comprised the mastoid air cells lined by cuboidal cells. On the outer aspect of the cortical bone and continuous with the outer layer of the peristeum, there was a fibro-osseous lesion in which rounded and ovoid zones of ossification comprising a mixture of slightly cellular woven and coarse lamellar bone was present (Figs. 1C, D). Occasional osseous structures included mature lamellar bone surrounded and encompassed by less mature bone which had a mixed woven and coarse lamellar pattern. Focal ongoing remodeling with osteoblastic and osteoclastic activity was seen (Fig. 1E). In some regions, there was direct continuity between the collagen fibers of the woven bone in particular and the accompanying stroma. The stroma comprised bland spindled cells arranged in an interwoven fasciculated manner. Nuclear atypia was not present and mitotic activity was not seen. Scattered mast cells were noted (Fig. 1F and insert). Permeation into the subjacent cranial bone was not seen and the surface of the skull was mildly irregular, merging with the fibrous stroma through a calcified tidemark (Fig. 1C). Immuno peroxidase stains carried out by a standard protocol and including pancytokeratin, EMA, CEA, and S100, were negative. None of the stromal cells were decorated by MIB1 (Ki 67) (Table 1).

The exact nature of the lesion was not clear, however the combined radiologic and histologic features of a bland fibro-osseous lesion in a surface location prompted concern for a subtle low-grade parosteal osteosarcoma (POS) and reexcision of the area was carried out to ensure clear margins. At 12 months follow-up, the patient is well and there is no evidence of recurrence.

**Case 2**

The patient, a 66-year-old female, had a 10 years history of a slowly growing lump behind the ear. CT scans, carried out originally in 2002 and repeated in 2009, showed a surface-based expanded lesion with widespread central rounded foci of ossification and an incompletely ossified rim. The lesion was closely related to the occipito-mastoid suture. There was no intra-osseous or intracranial extension (Fig. 2A). There was some remodeling of the cortex deep to the lesion. No obvious progression was evident over the intervening 7 years. At surgery firm bony tissue was adherent to the skull surface and removed with an osteotome.

**Pathologic Examination**

The specimen consisted of an oval nodule of firm bony tissue 32 × 14 × 8 mm, the surface of which was smooth and white. Microscopically, the tissue was composed of a slightly sessile nodular fibro-osseous lesion within the stroma of which variably distributed rounded, ovoid, and somewhat bossellated osseous structures were embedded, most comprising coarse lamellar bone (Figs. 2B, C). In some of the osseous structures mature lamellar bone was present, which in some regions was surrounded and encompassed by less mature bone which had a mixed woven and coarse lamellar pattern. Direct collagen continuity between the immature nodular bone and the surrounding fibrous stroma was evident in some regions (Fig. 2D). Focal ongoing remodeling with osteoblastic and osteoclastic activity was seen. The stromal component comprised fibrous tissue with evenly distributed bland fibroblastic cells arranged in an interwoven and fasciculated manner (Fig. 2E). The stromal cells had small ovoid nuclei with even nuclear borders and small nucleoli, and mitotic activity was not seen. Hyperchromatism was not noted. Focal mild myxoid change was evident (Fig. 2F). At the perimeter of the lesion, the fibrous tissue blended with surrounding fibro-adipose tissue and scattered entrapped adipocytes were present. Immunoperoxidase stains carried out by a standard protocol and including pancytokeratin, EMA, CEA, and S100, were negative. None of the stromal cells were decorated by MIB1 (Ki 67) (Table 1).

Although the possibility of a subtle low-grade parosteal osteosarcoma was again mooted, the long clinical history and lack of progression over 7 years made this seem highly unlikely and prompted reconsideration of the diagnosis in specimen 1.

Consultation with the New York Bone Club and in particular, Prof Peter Bullough, was undertaken and ultimately, given the location, imaging features, and histologic findings of osseous structures comprising woven and coarse lamellar bone with a rounded and somewhat psammomatoïd appearance lying in a bland fibrous stroma, a diagnosis of “Protuberant Fibro-osseous lesion of the Temporal Bone” was proffered in both cases.

**DISCUSSION**

In 1999, 2 cases of a unique exophytic fibro-osseous lesion of the temporal bone composed of spherical islands of mixed lamellar and woven within a dense bland seeming fibrous connective tissue stroma was documented. One was an 18-year-old male, the other a 21-year-old female. Both presented with a posterior auricular mass, one of 2 years duration the other noted 5 days before presentation. Both represented nontender exophytic masses without overlying soft tissue abnormalities. Imaging was...
FIGURE 2. A, Initial CT scan in 2002 shows the retroauricular mass with widespread central calcified foci and slight thinning and irregularity of the underlying cortex. No change in appearance was evident on a progress CT in 2009. B, At low power numerous variably sized round, ovoid and somewhat bosselated osseous structures lie within a fibrous stroma. C, On higher power the coarse lamellar and woven architecture is accentuated on polarisation microscopy. D, Polarisation microscopy shows occasional osseous structures in which more mature lamellar bone is present centrally bordered by woven bone. Focally direct continuity of collagen from the woven bone into the fibrous stroma is evident (arrow). E, The sparsely cellular stroma, containing a small round ossified structure, comprises interwoven fascicles of fibroblastic cells. F, In some regions the stroma has a more myxoid quality with interwoven collagen bundles.
similar representing a heterogeneous ossified mass emanating from the cortex of the temporal bone near the occipito-mastoid suture line on CT.

Both were excised using an osteotome. Involvement of the mastoid air cells was not seen.4-6 Follow-up at 15 and 17 months respectively revealed no recurrence. Subsequent follow-up 10 years later, before the publication of this paper, confirms that there has been no recurrence (personal communication with Prof Michael Klein). The lesions were classified as “Protuberant Fibro-osseous lesion of the Temporal Bone.”

The features in our cases have strikingly similar clinical, radiologic, and pathologic findings. The characteristic findings are those of a mass in the soft tissues of the retro-auricular region overlying the mastoid bone in which irregular dense matrix production is present, lying on the outer table of the skull, and confined to the surface of the bone. The lesions are reasonably well circumscribed without evidence of intramedullary or intracranial extension. Histologically, the lesion is confined to the soft tissues and comprises rounded and ovoid zones of woven and coarse lamellar bone within a bland fibrous stroma. In some regions direct continuity between the collagen fibres of the woven bone in particular and the accompanying stroma is present. The spindle/fibroblastic cells are arranged in a somewhat fasciculated manner. Nuclei are largely ovoid in contour with regular nuclear membranes and inconspicuous nucleoli. Nuclear atypia is not present and mitotic activity is not seen. The soft tissue margin of the lesional tissue is slightly irregular with some entrapment of fat although overall it is well defined.

Occasional foci of bone had central lamellar bone surrounded by woven bone prompting some concern regarding a “bone encasement” pattern. In an intraosseous location, this can be regarded as evidence of host bone permeation, a feature of malignancy.19 However, in this lesion the lamellar bone does not reflect original host bone as the lesion is entirely in soft tissue, thus its presence cannot be accorded the same significance.

Surface matrix producing lesions of bone always prompts consideration of a variety of benign tumors, reactive processes, and less frequently malignant tumors. The commonest exophytic or surface benign bone forming lesion in the skull is osteoma, which can be recognized radiologically as a dense mature sclerotic lesion on the surface of bone, often with a bossellated contour.6 It has been documented in the temporal bone.41 Histologically, it comprises dense compact bone with little stroma, which, if present comprises loose fibrovascular stroma.6,11,41 The other benign bone tumors, osteoid osteoma, and osteoblastoma, are rare in the skull, usually intraosseous in location and histologically comprise immature trabeculae of osteoid in a richly vascularised stroma with abundant plump osteoblastic rimming and numerous osteoclasts. A fibrous stroma is not expected.6,11 A rare case of osteoid osteoma is reported in the mastoid and periosteal osteoblastomas of the calvarium are documented.22,43

Surface cartilage containing lesions include periosteal chondroma, soft tissue chondroma, chondromyxoid fibromas, and periosteal chondrosarcomas, all of which are extremely rare in the calvarium.4-5,31 As fibro-osseous areas are not expected in these lesions and as neither lobulated hyaline cartilage (chondroma), immature lobulated myxochondroid stroma with intervening cellular areas (chondromyxoid fibroma) nor atypical chondro-osseous areas (periosteal osteosarcoma) are seen, these lesions can be excluded from consideration in the histologic differential diagnosis.

Surface reactive lesions including florid reactive periostitis, bizarre parosteal osteochondromatous proliferation, turret and subungual exostoses, and fibro-osseous pseudotumor, usually occur in acral locations, have a clinical history of rapid evolution and often, although not inevitably, have a history of prior trauma.32 Rare cases involving the skull are documented.25,30

Florid reactive periostitis is characterized by trabeculae of immature woven osteoid, lined by plump osteoblasts within a cellular fibroblastic stroma resembling nodular fasciitis with plump and stellate fibroblasts randomly oriented or arranged in short fascicles. Intermingled red cells are common and mitoses frequent. In bizarre parosteal osteochondromatous proliferation, irregular areas of calcifying cartilage with a purple tinctorial quality are seen, the stroma is cellular with plump fibroblasts and immature bone trabeculae lined by plump osteoblasts are usually present. Turret and subungual exostoses have a more orderly maturation with immature woven bone bordered by osteoblasts at the periphery merging with increasingly mature bone with a more lamellar architecture but in a vascularised stroma at the base, close to the bone surface.14,25,30,32,36 Reactive surface lesions of larger bones are rare and include myositis ossificans, which may reside close to bone stimulating a periosteal response simulating a true surface lesion.14 In most reactive lesions, in particular myositis ossificans, characteristic zonation of the components is present with the most mature ossification peripherally, not evident in our cases.6,14

These features contrast with the essentially inactive appearance of the fibrous stroma, relative lack of osteoblastic activity, and lack of any cartilaginous component seen in our cases. In none of these entities is a bland paucicellular fibrous stroma expected and rounded orderly zones of ossification are not seen.

The commonest fibro-osseous lesions of the craniofacial bones, including that of the temporal bone, are fibrous dysplasia and ossifying fibroma. These lesions are always centered in an intraosseous location.44 Although protuberant forms of fibrous dysplasia have been documented (so called “Fibrous Dysplasia Protuberans”), these lesions are exceptional, have not been reported in the skull and all have had a confirmed intraosseous component in combination with the exophytic area.10

Surface fibro-osseous lesions of bone without an intraosseous component are rare and usually classified as parosteal osteosarcoma.13,39 POS, also known as juxtapercortical osteosarcoma, is a rare slow growing surface bone tumor accounting for approximately 4% to 6% of
all osteosarcomas. It usually presents as a sclerotic dense sessile mass on the surface of bone, and there is often a cleft between the tumor and the underlying cortex on both plain radiograph and CT, reflecting its’ origin from the outer layers of the peristium. Distinctly rounded osteosarcomas are rare. In general, the bone trabeculae have a parallel arrangement within the fibrous stroma and at the periphery irregular infiltration of the surrounding soft tissues is common. There may be a peripheral area of chondroid tissue resembling a cartilage cap, of which 5 involved the mastoid bone.

In 2 of these, there was a history of antecedent trauma neither of which is illustrated histologically although 1 shows similar imaging features to our cases. One patient had a 20 years history without documented change in size for 9 years and the remaining 2 cases had a 1 and 3 year history, respectively. All 3 had a dimension of 3 to 5 cm. One was reviewed by 3 eminent bone pathologists and characterized as POS grade 2, suggesting a degree of atypia, not seen in our cases.

The pathogenesis of protuberant fibro-osseous lesion of the temporal bone is elusive. The mastoid bone forms the posterior part of the temporal bone and is formed by intramembranous ossification in the maxillary prominence of the first branchial or pharyngeal arch, which subsequently forms the squamous temporal, maxillary, and zygomatic bones. The middle ear develops from the tubotympanic recess of the first pharyngeal pouch, the distal component of which expands and becomes the tympanic cavity. During the late fetal period expansion of the tympanic cavity gives rise to the mastoid antrum located in the petromastoid part of the temporal bone. By 2 years of age the mastoid air cells are well developed and produce conical projections of the temporal bone known as the mastoid process.

The outer surface of the mastoid is rough, giving attachment to the mastoid fascia, occipital belly of the occipito-frontalis, and the auricularis posterior. The lateral surface of the mastoid process gives insertion to the sternocleidomastoid, splenius capitis, and longissimus capitis. On its medial side, there is a deep groove termed the mastoid notch for the attachment of the posterior belly of the digastric and medial to this notch the shallow occipital groove lodges the occipital artery. In all cases documented thus far, the lesions are closely related to the occipito-mastoid suture and as such, the possibility of a reactive lesion with membranous bone formation related to trauma in particular must be considered. On direct questioning, there was no evidence of prior trauma, both patients had a symmetrical facial and skull appearance and posture and occupational and recreational factors did not seem significant. Unilateral involvement would be unlikely in this location in the absence of a specific precipitant. Histologically, features to suggest active trauma including granulation tissue, haemosiderin deposition, organising fibrosis, xanthomatous change, or fasciitis like stroma were not seen. A zonal pattern, expected in posttraumatic fibro-osseous lesions of bone (most often identified in the rib), was not present. In avulsive cortical irregularity/perioseal desmoid, irregularity of the cortex is usual on imaging and histologically, fibromatosis-like changes are expected. Bone formation, as seen in these cases, is not expected. Remodeling of the cortical surface with irregularity confirmed histologically in case 1 would support the likelihood of a subtle reactive process, possibly related to remote repetitive trauma.

The possibility of a lesion of meningeal origin, particularly in view of the somewhat psammomatoid appearance of the osseous component has been considered, however, the affected tissue is sufficiently removed from the meninges that in the absence of an unusual meningeal rest this seems unlikely, particularly given the lack of any corroborative evidence on H&E and epithelial membrane antigen is negative.

The long clinical history and imaging follow-up documented in our Case No 2 supports interpretation as a benign lesion. The lack of documented recurrence in the earlier cases, absence of parallel trabecular architecture of the osseous component, sole presence of rounded and ovoid osseous structures, and lack of nuclear atypia allows distinction from low-grade parosteal osteosarcoma.

We believe that this is a specific and recognizable lesion, which is confined to the soft tissues of the surface of the temporal bone/mastoid air cell region with distinctive and unique imaging and pathologic findings.

We would suggest that in the appropriate clinical and imaging setting, a conservative approach to management be adopted. Local excision seems to be adequate.

As both of these cases were kindly reviewed by the Members of the New York Bone Club and recognized solely by Professor Peter Bullough, who was the pathologist involved in the original description of this lesion, rather than using the somewhat cumbersome term “protuberant fibro-osseous lesion of the temporal bone,” although descriptive, we would suggest the more succinct term of “Bullough lesion” or even “Bullough Bump!”

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