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Case Report

Fibro-osseous lesion of the cranium in an adolescent patient

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Abstract

Background: Fibrous dysplasia, ossifying fibroma, and desmoplastic fibroma are rare benign calvarial lesions, which can have local aggressive behavior. These tumors can present with similar clinical and radiologic characteristics making diagnosis difficult at times.

Case Description: A 16-year-old male presents after noting an indentation of his skull. Comparison with current and previous imaging revealed progressive erosion of the skull underlying the indentation.

Conclusion: Fibrous dysplasia, ossifying fibroma, and desmoplastic fibroma are rare fibro-osseous tumors with similar characteristics radiographically. Accurate diagnosis of these tumors can be difficult even with the combination of clinical presentation, imaging, and pathology. The treatment of choice is resection and cranial reconstruction, if necessary, with close follow-up as recurrence can occur.

Key Words: Desmoplastic fibroma, fibrous dysplasia, intraosseous, ossifying fibroma, skull lesion

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INTRODUCTION

Tumors of the skull are uncommon and account for 1–4% of all bone tumors.[27] Fibrous dysplasia (FD), ossifying fibroma (OF) (aka osteofibrous dysplasia outside of the head and neck region), and desmoplastic fibroma (DF) are benign fibro-osseous tumors in the World Health Organization (WHO) classification of tumors affecting the cranium.[10] However, given that these tumors can present with very similar features diagnosis can be difficult at times. Therefore, the clinical presentation, imaging studies, and histological characteristics should always be assessed together. Here we present a 16-year-old male with a bony lesion of the right parietal skull with conflicting imaging and pathologic findings.

CASE REPORT

Presentation

A 16-year-old male presented for evaluation of an indentation of his right parietal skull. The patient had first noted the indentation 2 weeks prior with no recent change in size. He had been involved in a 4-wheeler accident 2 years before and was evaluated for a possible concussion during a football game about one and a half years prior to presentation. The patient described having

occasional headaches, but had no local tenderness or other neurological symptoms.

Examination

On examination, the patient was noted to have a palpable defect and indentation of the right parietal bone. The overlying scalp was unremarkable and he was neurologically intact.

Imaging

The patient's previous imaging was reviewed. The first study, from 2 years prior, showed a skull defect in the right parietal bone [Figure 1a]. The second and third computed tomography (CT) scans showed progressive erosion of the skull [Figure 1b and c]. The defect did not appear to involve the dura or scalp. A magnetic resonance imaging (MRI) was obtained and confirmed the localization of the lesion to the skull [Figure 1e-f].

Management

Excision of the skull defect and right parietal cranioplasty with titanium plate was performed. The overlying tissue was grossly abnormal, measuring approximately 5 cm in diameter, and was removed en bloc. The underlying bone was thin and eggshell like with a trabeculated pattern, especially in the center. Peripherally, the bone was thickened with a sponge-like appearance. The abnormal tissue portion could be seen replacing the diploic space while leaving the inner and outer table intact at the periphery. The dura was not involved and was stripped from the bone and left intact. The abnormal bone was then removed and a mesh titanium plate was used to replace the cranial defect [Figures 1d and 3].

The patient tolerated the operation well and had no immediate operative or perioperative complications. The right parietal indentation was corrected and the patient was discharged on postoperative day 2.

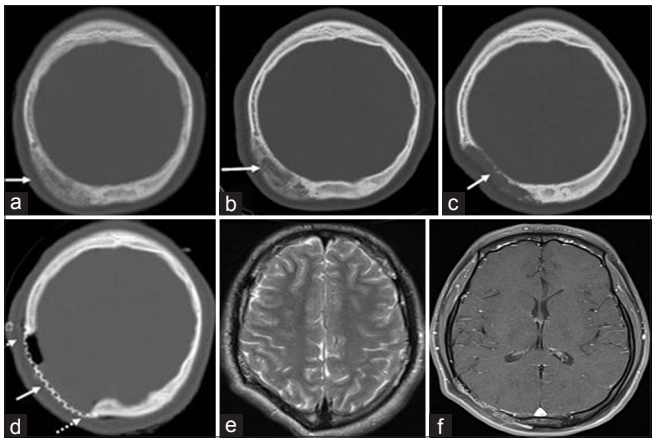

Figure 1: CT scan of the brain from March 16, 2012 (a) shows bony lesion with predominant ground glass appearance which is extracranial.Additional CT on September 14, 2012 (b) and February 21, 2014 (c) shows progression of the bony lesion with increase in the area of lucency. (d) post-op CT showing reconstruction with titanium mesh. (E-f) pre-operative showing extramedullary lesion with dural enhancement

Pathology

On gross examination, the soft tissue lesion appeared rubbery, yellow, and avascular. The tissue consisted predominantly of dense fibrous tissue with a few foci of woven bone at the periphery adjacent to the eroded bone. Angulated spicules of bone were rare, some of which lacked osteoblastic rimming while others were rimmed. There was minimal inflammation. The final diagnosis was that of a benign fibro-osseous lesion most in keeping with a DF. The case was reviewed at the Mayo Clinic where it was felt to be an atypical FD. Conflicting pathology interpretations of these lesions is not uncommon.

DISCUSSION

Clinical

FD accounts for 2.5% of benign bone tumors and 7% of all bone tumors.[29] FD is defined by WHO as a "benign medullary fibro-osseous lesion which may involve one or more bones."[10] It tends to be slow growing, but is chronically progressive with the gradual replacement of bone with fibrous connective tissue. Children and adolescents are more often affected than adults.[29] The mandible is most commonly affected,[10] but any bone can be involved with a greater tendency for lesion formation in the long bones and ribs in women and in the skull in men. Overall there is equal distribution between both sexes. FD of the skull can affect any of the cranial bones, but most commonly affects the ethmoid and frontal bones typically presenting with cranial asymmetry.[29]

FD can present as monostotic, polyostotic, or McCune–Albright syndrome subtypes. The monostotic subtype is six times more common. FD involving the cranium tends to be monostotic in nature. FD involving the femur, tibia, pelvis, and spine tend to be polyostotic. Polyostotic FD is often associated with McCune–Albright syndrome and tends to be more severe with an earlier onset. McCune–Albright syndrome is characterized by Albright's triad of early onset of puberty in females, cutaneous hyperpigmentation, and endocrinopathies. A somatic mutation in the G-protein alpha subunit gene GNAS1 may be responsible for causing FD.[10,29]

OF is a slowly growing benign tumor that is locally aggressive with a high recurrence rate. It tends to occur mostly in the third and fourth decades of life. Both sexes are affected equally. Aggressive behavior can occur in young patients with a gradual decrease in aggressiveness with age. OF affects the head and neck region and is analogous to osteofibrous dysplasia listed in the WHO classification, which mainly affects the tibia. OF is most often found in the mandible and maxilla and is less commonly found in the nasal bones, ethmoid cells,

orbit, and cranium. Typical patients present with cranial asymmetry and swelling.[10,28]

DF is rare, making up 0.3% of benign bone tumors and 0.06% of all bone neoplasms.[24] DF was first described by Jaffe *et al.* in 1958 and is defined by WHO as a "rare, benign bone tumor, composed of spindle cells with minimal cytological atypia and abundant collagen production.[10] Although histologically benign with a slow growth rate, it is locally aggressive with a high rate of recurrence.[5,10] DF is typically seen in both sexes with similar frequencies and a predilection for patients aged less than 30 years.[7,13] It usually affects the metaphysis of long bones, mandible, and pelvis, while it less commonly involves the maxilla, sternum, and vertebrae.[5,24]

Gardini *et al.* first described a case of DF affecting the skull in 1978 and a total of 20 cases have been reported[2,6-9,11,12,15,17,18,21-24,26,31,32] with only 8 of these involving pediatric patients.[2,7,11,22,31,32] DF of the skull tends to have a higher incidence in females.[7,18,24] DF typically presents with cranial asymmetry and headaches. Additionally, DF of the skull can involve the dura mater.

Imaging

Radiographically, FD has a sclerotic, lytic, or pseudopagetic appearance. There is a characteristic ground glass matrix in the sclerotic variant, which is the most common type, making up 50% of FD. The lytic form is characterized by radiolucency, while the pseudopagetic variant is a combination of the sclerotic and lytic forms. FD is nonaggressive and often localized between diploe. There is normally no periosteal reaction or soft tissue extension. CT scans are particularly helpful in visualizing the ground glass pattern [Figure 2e and f], while MRI can provide additional information such as cranial nerve and dural involvement [Figure 2d and g].[10,29]

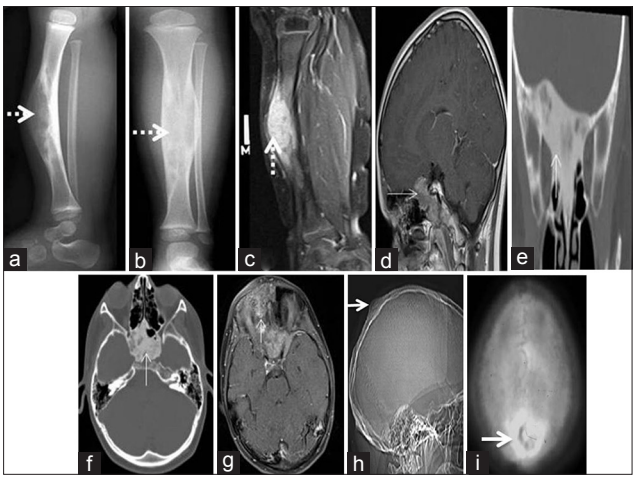

Figure 2: Lateral (a) and AP (b) X-ray showing an osteofibrous dysplasia of the tibia (*broken arrow*). Sagittal postcontrast MRI (c) using enhancement of the tibial lesion (*broken arrow*). This lesion is synonymous to ossifying fibroma and is name based on its location. Sagittal (d) and axial (g) postcontrast MRI showing fibrous dysplasia (*thin arrow*). Coronal (e) and axial (f) CT scan showing fibrous dysplasia (*thin arrow*). Lateral skull X-ray (h) and axial CT (i) of a desmoplastic fibroma[8]

OF is a radiolucent, well demarcated round or oval mass with thinning cortical bone boundaries resembling an eggshell. Older lesions are visibly calcified. CT scans show a cortical epicenter to the lesion that is separated from medullary bone by sclerosis and does not invade the soft tissue [Figure 2a and b]. MRI shows mixed signals in T1-weighted images and high intensity signals in T2-weighted images with enhancement on contrast [Figure 2c].[10,28]

DF is osteolytic with expansion of the bone with the original cortex being replaced by a thin shell of new bone. The lytic area has a trabeculated or bubble-like appearance.[5] CT scans are particularly useful in order to identify the degree of local bone destruction[24] [Figure 2i]. The degree of involvement of the scan varies from cortical thinning to complete destruction of the cortex.[7,14,23] MRI findings are not completely characterized in the literature, but can help in confirming displacement of local soft tissue without any clear signs of local infiltration.[14,24] The hypocellularity and dense collagenous tissue of DF results in intermediate signal intensity in T1-weighted images and a heterogeneous intensity in T2-weighted images.[5]

Although there are radiographic characteristics to look for that can help in differentiating these tumors, there are similarities that make this difficult and correlation with pathological evaluation is crucial.[29]

Pathology

Grossly, FD is tan to gray or white with a gritty, firm texture. Cysts filled with yellowish fluid can be found. Histologically, FD tends to be well circumscribed with fibrous and osseous components. The fibrous component has bland spindle-shaped cells with a low mitotic rate and the osseous component has curvilinear trabeculae of bone lacking osteoblastic rimming, an important diagnostic feature. Foamy cells, multinucleate giant cells, secondary aneurysmal bone cyst, or myxoid change may be present [Figure 4e-h].

OF is grossly white to yellow, or red in color and soft or gritty in texture. The cortex is thinned, but periosteum is intact. There is usually a sclerotic rim. OF shows irregular fragments or spicules of woven bone with osteoblastic rimming of lamellar bone with occasional osteoclasts. The fibrous component consists of bland spindle cells. The center of the lesion consists mainly of the spicules of bone and the peripheral lamellar bone often blends with the normal surrounding bone [Figure 4i and j].

DF is grossly white to tan in color with a rubbery texture. Histologically it is hypocellular composed of fibroblasts

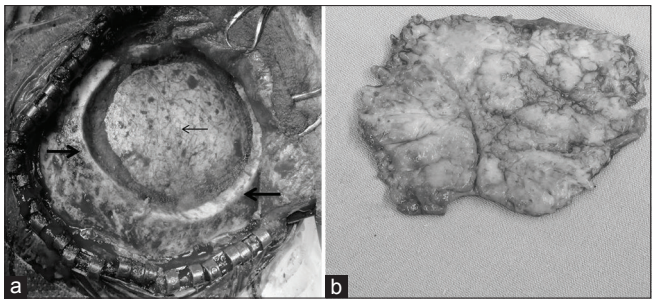

Figure 3: (a) Shows intraoperative image of craniectomy with good margins (*large arrows*). There is no involvement of the dura mater (*thin arrow*). (b) Shows the resected cranial defect

with ovoid to spindled nuclei in a variably hyalinized collagenous matrix [Figure 4a-d].[24] DF can extend into surrounding bone and may eventually expand into the surrounding soft tissue as seen in this case.[5,16]

Management

Treatment of skull FD involves surgical removal of the lesion. Total resection with craniofacial or cranial reconstruction is the preferred method of treatment, although nerve function and aesthetics should be taken into consideration. This yields a good prognosis with a low rate of recurrence. In a retrospective study following 81 patients with FD of the skull, 13.7% of the follow-up patients with complete excision had tumor recurrence with 0–2 cm variance in margin widths.[29] Malignant transformation of FD is rare but repeated follow-up is recommended.[10,29]

The locally aggressive nature of OF makes surgical resection the preferred treatment. Despite this, when OF involves areas around vital structures, periodic removal of the lesion and preservation of the vital structures with follow-up is warranted. Reported rates of recurrence vary from 0% to 28% after total primary resection and are higher in subtotal resections.[30] In a case report of five patients with OF, four who underwent complete resection showed no recurrence and one who underwent partial resection showed recurrence of tumor growth.[1]

The treatment of choice for DF of the skull is complete resection, with tumor-free margins due to the locally aggressive and infiltrative nature of DF.[24] The involvement of the dura in certain cases warrants removal of the affected dura with duraplasty. Cases of DF of bone other than the skull are reported to have recurrence rates of 17–30%.[16] Zero percent of the previously reported cases of DF of the cranium have recurred. However, continued follow-up with imaging is warranted to monitor for possible recurrence given that these lesions are reported to recur at other sites.

In our patient, an en bloc resection of the affected portion of the skull was performed, while leaving the uninvolved dura intact [Figure 3a and b]. Cranioplasty with a titanium plate was used to reconstruct the

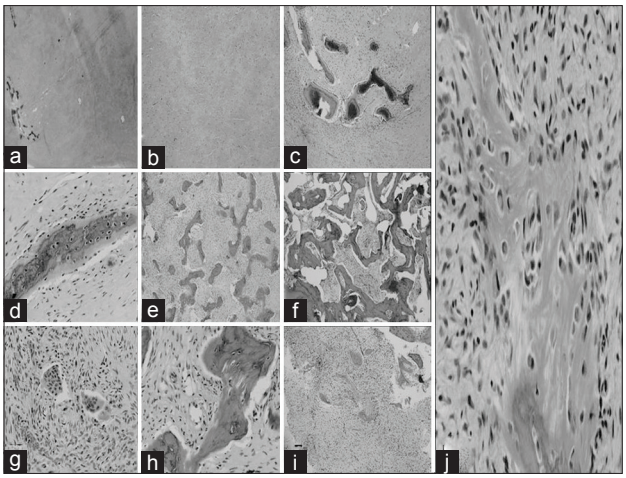

Figure 4: Desmoplastic fibroma. (a) Dense fibrous tissue with spicules of woven and laminated bone at periphery. (b) Uniform spindle cells in the stroma. (c) Laminated bone at periphery with osteoblastic rimming. (d) Woven bone with osteoblastic rimming. Fibrous dysplasia. (e) fibrous stroma with angulated spicules of bone. (f) Spicules of bone with no osteoblastic rimming. (g) Occasionally osteoclasts can be seen. (h) Spicule of bone without osteoblastic rimming. Ossifying fibroma. (i) Cellular stroma with spicules of bone. (j) Spicule of bone with osteoblastic rimming

skull [Figure 1d]. The recommended treatment of choice for FD, OF, and DF of the skull is surgical resection with cranial reconstruction as necessary followed by regular follow up. In cases of recurrence, effective chemotherapy and radiotherapy have not yet been clearly established.

Adjuvant therapies

Nonsurgical management of FD is not effective.[4] In a series of FDs with secondary aneurysmal bone cysts, adjuvant radiotherapy led to the development of sarcoma resulting in the abandonment of this approach.[19] The limited number of reported cranial cases of OF have been described as being treated surgically.[30] While there may be a role for radiotherapy in DF there is limited literature on this modality of treatment.[20,25] The mainstay of treatment in these patients remains surgical resection, although radiation can be considered when surgical treatment is not an option.[20,25] There have been reports of one-third of desmoid tumors, with similar morphologic and behavioral characteristics to DF, being positive for estrogen receptors and subsequently responding to hormonal therapy.[3] However, the reported cases of DF of the skull have have not been shown to be positive for estrogen receptors.[8] The role of adjuvant radiotherapy and chemotherapy remains largely un-established in these tumors.

CONCLUSION

FD, OF, and DF of the skull are rare and often present with somewhat similar clinical and radiographic features. In our case, the radiological diagnosis was that of an OF and the

initial pathology was that of a fibro-osseous lesion most in keeping with a DF. Review at another institution suggested an atypical FD. Our case highlights that the difficulty in diagnosing these bony tumors of the skull. Diagnosis requires consideration of the clinical, radiographic, and pathology. The preferred treatment for all three involves en bloc surgical resection due to potential locally aggressive behavior. Regular follow up is strongly recommended.

REFERENCES

1. Artico M, Cervoni L, Salvati M, Raco A, Ciappetta P. Ossifying fibroma of the skull: Clinical and therapeutic study. Tumori 1994;80:64-7.
2. Bahri I, Kallel R, Gouiaa N, Boudawara Z, Boudawara TS. Desmoplastic fibroma of the skull in an infant. Neurochirurgie 2011;57:39-41.
3. Brooks MD, Ebbs SR, Colletta AA, Baum M. Desmoid tumours treated with triphenylethylenes. Eur J Cancer 1992;28A: 1014-8.
4. Cai M, Ma L, Xu G, Gruen P, Li J, Yang M, *et al.* Clinical and radiological observation in a surgical series of 36 cases of fibrous dysplasia of the skull. Clin Neurol Neurosurg 2012;114 :254-9.
5. Campanacci M, Ruggieri P, Gasbarrini A, Ferraro A, Campanacci L. Osteoid osteoma. Direct visual identification and intralesional excision of the nidus with minimal removal of bone. J Bone Joint Surg Br 1999;81:814-20.
6. Celli P, Cervoni L, Trillo G. Desmoplastic fibroma of the skull. Case report and review of the literature. Neurochirurgie 1997;43:260-4.
7. Cho BH, Tye GW, Fuller CE, Rhodes JL. Desmoplastic fibroma of the pediatric cranium: Case report and review of the literature. Childs Nerv Syst 2013;29:2311-5.
8. Deniz K, Ceylan D. Desmoplastic fibroma of the skull. Acta Neurochir 2008;150:285-90.
9. Dutt SN, Mirza S, Irving RM, Jones EL. Desmoplastic fibroma of the temporal bone. J Laryngol Otol 2000;114 :314-7.
10. Fletcher CD, Unni KK, Mertens F. Pathology and genetics of tumours of soft tissue and bone. Lyon: IARC Press; Oxford: Oxford University Press; 2002.
11. Gardini G, Minghetti G, Ferracini R. Desmoplastic fibroma of frontal bone. Report of a case (author's transl). Pathologica 1978;70 :575-9.
12. Goldberg AN, Janecka IP, Sekhar LN. Desmoplastic fibroma of the skull: A case report. Otolaryngol Head Neck Surg 1995;112:589-91.
13. Greenspan A, Remagen W. Differential diagnosis of tumors and tumor-like lesions of bones and joints. Philadelphia: Lippincott-Raven; 1998.
14. Haney J, Olson PN, Griffiths HJ. Radiologic case study. The clinical and radiologic features of desmoplastic fibroma of bone. Orthopedics 1994;17:77:80-75, 88.
15. Hufnagel TJ, Artiles C, Piepmeier J, Kier L, Kim JH. Desmoplastic fibroma of parietal bone simulating eosinophilic granuloma. Case report. J Neurosurg 1987;67:449-51.
16. Inwards CY, Unni KK, Beabout JW, Sim FH. Desmoplastic fibroma of bone. Cancer 1991;68:1978-83.
17. Lath R, Ranjan A, Ratnam BG, Gowrishankar S. Desmoplastic fibroma of the frontal bone. Neurol India 2006;54:314-5.
18. Lee S, Jung SM, Cho BK, Kim H. Desmoplastic fibroma of the cranium in a young man. J Korean Neurosurg Soc 2012;52:561-3.
19. Manjila S, Zender CA, Weaver J, Rodgers M, Cohen AR. Aneurysmal bone cyst within fibrous dysplasia of the anterior skull base: Continued intracranial extension after endoscopic resections requiring craniofacial approach with free tissue transfer reconstruction. Childs Nerv Syst 2013;29:1183-92.
20. Nag HL, Kumar R, Bhan S, Awasthy BS, Julka PK, Ray RR. Radiotherapy for desmoplastic fibroma of bone: A case report. J Orthop Surg (Hong Kong) 2003;11:90-3.
21. Okuno H, Yamamichi N, Watanabe I, Tokita N. Desmoplastic fibroma of the temporal bone. J Otolaryngol 1990;19:311-4.
22. Ovul I, Oztop F, Zileli M. Congenital desmoplastic fibroma of the cranium. Childs Nerv Syst 1988;4:45-6.
23. Pensak ML, Nestok BR, Van Loveren H, Shumrick KA. Desmoplastic fibroma of the temporal bone. Am J Otol 1997;18:627-31.
24. Rabin D, Ang LC, Megyesi J, Lee DH, Duggal N. Desmoplastic fibroma of the cranium: Case report and review of the literature. Neurosurgery 2003;52:950-4.
25. Sanfilippo NJ, Wang GJ, Larner JM. Desmoplastic fibroma: A role for radiotherapy? South Med J 1995;88:1267-9.
26. Selfa-Moreno S, Arana-Fernandez E, Fernandez-Latorre F, Diaz-Ramon C, Ricart-Rodrigo M, Moreno-Flores A. Desmoplastic fibroma of the skull. Case report. Journal of neurosurgery 1995;82:119-20.
27. Sutherland JE, Dahdaleh NS, Greenlee JD, Kirby PA, Sargent MD. Large osteolytic skull tumor presenting as a small subcutaneous scalp lesion. J Am Board Fam Med 2011;24:763-7.
28. Vlachou S, Terzakis G, Doundoulakis G, Barbati C, Papazoglou G. Ossifying fibroma of the temporal bone. J Laryngol Otol 2001;115:654-6.
29. Wei YT, Jiang S, Cen Y. Fibrous dysplasia of skull. J Craniofac Surg 2010;21:538-42.
30. Wilson JR, Kumar R, Goddard A, Liddington M, Carter L, Russell J, *et al.* Recurrent ossifying fibroma of the sphenoid bone 26 years after primary surgical excision; A case report and review of the literature. Acta Neurochir (Wien) 2013;155:549-51.
31. Wolfe SQ, Cervantes L, Olavarria G, Brathwaite C, Ragheb J, Morrison G. Desmoplastic fibroma of the pediatric skull. Report of three cases. J Neurosurg 2005;103 (4 Suppl):362-5.
32. Yoon SH, Kim SH, Shin YS, Ahn YW, Cho KG, Lee KB, *et al.* Desmoplastic fibroma of the skull in an infant. Child's nervous system 2006;22:176-81.