Romanian Neurosurgery | Volume XXX | Number 4 | 2016 | October - December

Article

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INDIA

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DOI: 10.1515/romneu-2016-0088

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Giant osteoma of the skull vault: A rare case of mixed variety

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Abstract: Osteoma is the most common primary bone tumor in the craniofacial skeleton. However, most of these are small, asymptomatic and arise from the facial bones or in relation to the paranasal sinuses. Cranial vault osteomas, that too giant and symptomatic are much rarer. We report a case of sixty year-old gentleman presented with a very slowly increasing, painless, hard swelling on the left side of his head. Computerized tomography scan showed the left parietal calvarial tumor to be having large exostotic and enostotic components. He underwent an en-bloc excision of the tumor and cranioplasty. Giant, symptomatic cranial vault osteoma with concurrent exostotic and enostotic components is extremely rare. These lesions can be safely and completely excised with careful planning and attention to detail.

Key words: Cranial osteoma excision, enostotic, exostotic, giant osteoma, skull tumor

Introduction

Osteoma is the most common primary bone tumor in the craniofacial skeleton, reported to affect 0.43% of the population. (7) However, most are small and asymptomatic, arising in the paranasal sinuses and facial bones. Cranial vault osteomas, that too giant and symptomatic are much rarer. Even when found, these tumors have been described to grow either extracranially (exostotic) or intracranially (enostotic). A tumor with both exostotic and enostotic components has not been described so far. Such a tumor poses a unique set of challenges for surgical management. We report the first case of a giant, symptomatic cranial vault osteoid osteoma of mixed variety (with giant exostotic and enostotic components) and discuss the nuances of its safe and complete surgical excision.

Case report

A sixty year-old gentleman presented with a very slowly increasing, painless, hard swelling over the left parietal region over the past 10 years. The swelling had now attained a very large size and he had holocranial headache over the last 2 months and a feeling of clumsiness and fatigability in the right hand. He did not have swellings anywhere else in the

body, nor did any of his family members have similar complaints. On examination, a large swelling was noted over the left parietal region, measuring about 16cmx13cm, having a smooth surface. The swelling extended from just behind the coronal suture to the iniomastoid line anteroposteriorly and from 2 cm off the midline to 2 cm short of the base of the mastoid mediolaterally. (Figures 1 and 2) It was nontender and bony hard to palpation. His neurological examination revealed papilloedema and a pronator drift in the right upper limb. The deep tendon reflexes were brisk on the right side. A computerized tomography (CT) scan showed the lesion to be having a large enostotic component, in addition to the clinically obvious exostotic component. The enostotic component was extending further more anterior and medial than the limits of exostotic component. The former was causing mass effect on the underlying brain, resulting in a midline shift of >10mm to the right. (Figure 3) A diagnosis of giant left parietal osteoma was made. He underwent complete excision of the tumor and cranioplasty with polymethyl methacrylate. A large left temporo-parieto-occipital square-scalp flap based inferiorly was raised, leaving the pericranium on the tumor. Several burr holes were placed all around the tumor and on either side of the sinuses to be exposed during craniotomy. After careful separation of underlying dura and sinuses, burr holes were connected with craniotome. The free bone flap containing the entire tumor was elevated while dissecting the underlying dura adherent to the enostotic component with fine periosteal elevators. The dissection was started anteriorly, progressing medially and posteriorly. The superior sagittal sinus, left transverse sinus and finally the torcula were carefully separated and the entire tumor, along with a margin of >1cm was excised en-bloc (Figure 4). There was no dural breach by the tumor. The venous ooze from the exposed sinuses was controlled with gelfoam and the bleeding from dural vessels controlled by generous use of bipolar coagulation. Cranioplasty was performed with acrylic, placing multiple dural hitch stitches. The histology of resected tumor confirmed features of osteoid osteoma. Wound drain was removed after 5 days. But he developed subgaleal collection under the skin flap which was treated with aspiration, antibiotics and application of crepe bandage (Figure 5). Subsequently, the wound healed well. By three months, he had been relieved of headache, right hand weakness had completely improved and normal skull contour had returned. He continues to be asymptomatic at one year follow-up (Figure 6).

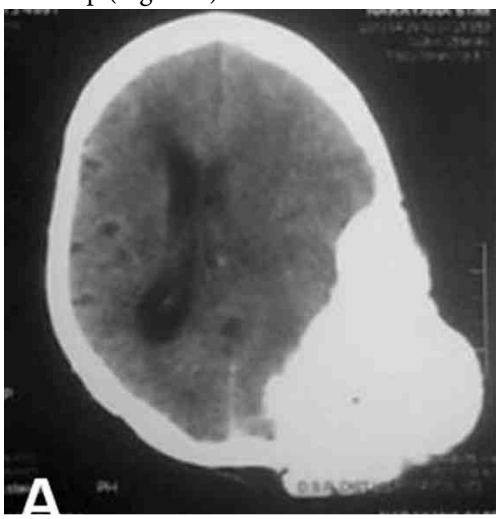

Figure 1

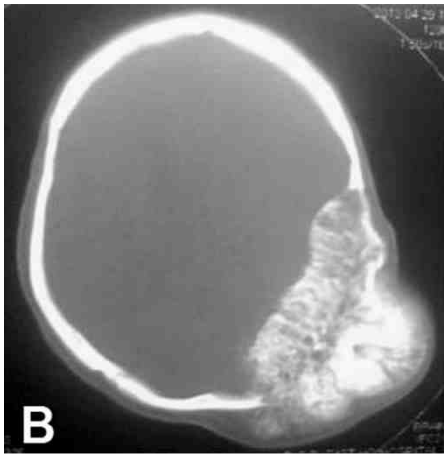

Figure 2

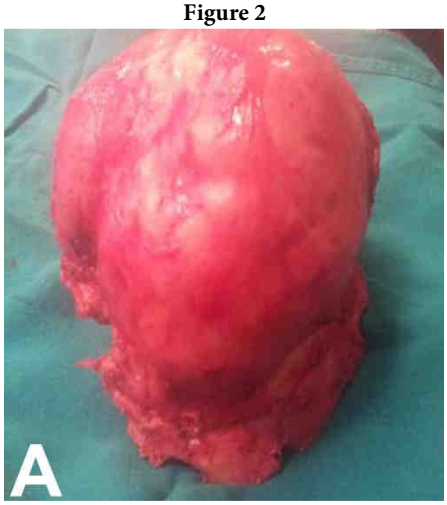

Figure 3

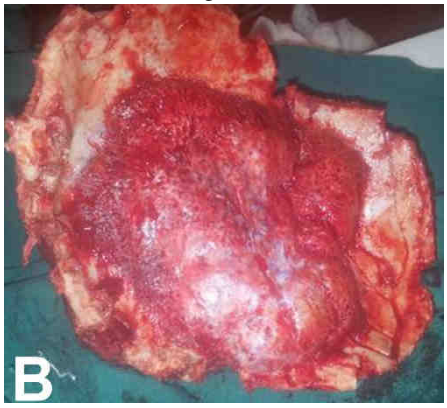

Figure 4

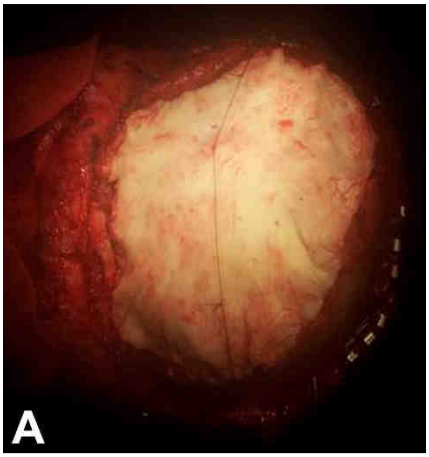

Figure 5

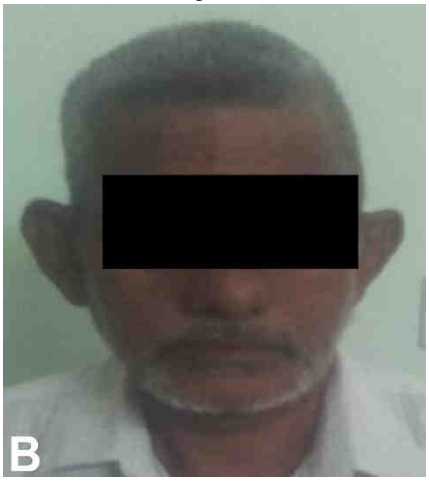

Figure 6

Discussion

Osteoma is the most common primary benign tumor in the craniofacial skeleton. A vast majority of these arise in relation to the paranasal sinuses or mandible. (7) Cranial osteomas have been classified by Haddad FS et al. into four types: 1 - intraparenchymal, 2 - dural, 3 - skull base and 4 - skull vault. The skull vault osteomas are further subdivided into two types: exostotic (arising from outer table) and enostotic (arising from inner table

and growing intracranially). (4) Although not frequently described in literature, exostotic osteomas are believed to be much more common but most often asymptomatic. (5) Enostotic and symptomatic osteomas are rare. To our knowledge, an osteoma having both exostotic and enostotic components has not been reported so far. This is the first such report with giant exostotic and enostotic components in a symptomatic cranial vault osteoma. The symptomatic tumors have been reported to present with headache, seizures, visual symptoms (when orbit is involved), pneumocephalus, mucocele and abscess (when paranasal sinus is involved) and even hydrocephalus due to superior sagittal sinus involvement. (2, 4, 5) Our patient presented with headache and right pyramidal signs probably due to direct compression of motor cortex by the enostotic component of tumor. Previously, the term "large" osteoma has been used for a tumor greater than 3cm and the term "giant" reserved for those larger than 6 cm. (5) The tumor in our patient was much larger, with the greatest dimension being 16cm. Only one skull vault osteoma previously described, has been slightly larger (17.5cm). (3) Surgical management of these benign tumors is most often simple and curative. However, management of the large ones with involvement of paranasal sinus and/or orbit is more complicated and has been well described by other workers. (1, 6) The giant tumors involving the vault, especially when containing both exostotic and enostotic components, present a different set of challenges. The planning of craniotomy requires careful surface marking of the enostotic component apart from the obvious exostotic component. Where facilities are available, use of neuronavigation at this stage can be helpful. After disconnecting the bone flap containing the tumor from the surrounding bone, care has to be taken to separate the dura from the enostotic component. A lesion as large as the one in our patient is likely to be overlying one or more venous sinuses and utmost care has to be exercised in dissecting the tumor from them. In our patient there was no dural breach. But lesions with mushroom-like growth can breach the dura and Haddad FS et al. have described disconnecting the overlying bone flap in three separate pieces before dissecting the enostotic tumor off the underlying brain and blood vessels. (4) During cranioplasty it is important to place multiple dural hitch stitches to obliterate the large epidural dead space. It is probably appropriate to excise the excess and redundant skin prior to closure. It was not done in our patient and led to persistent wound collection under the redundant skin flap, although eventually wound healed well without any major consequences.

Conclusion

Giant, symptomatic cranial vault osteoma with concurrent exostotic and enostotic components is very rare. A safe and total excision of these lesions is possible, with careful planning and attention to detail. Surgical excision provides complete cure, with excellent outcome.

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