Intradiploic meningioma of the orbit: A case report

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We report a case of a 12-year-old male child who presented with a gradual onset exophthalmos involving the left eye. The plain radiographs of the skull showed hyperostosis of the left orbital roof. The computerized tomography (CT) scan revealed an intradiploic orbital roof tumor with expansion of both the tables of the orbital roof. The tumor was completely excised by an extradural route using a basal frontal craniotomy. The histopathological diagnosis of the tumor was a psammomatous meningioma. A split calvarial frontal bone graft was used to reconstruct the orbital roof. The pathogenesis, radiological features and surgical technique involved in the management of intradiploic orbital roof meningioma are discussed and the relevant literature is reviewed.

**Key Words:** Intradiploic, meningioma, orbital roof

**Introduction**

Primary intraosseous meningiomas of the skull are rare tumors. They are also known as ectopic meningiomas. The intradiploic meningiomas of the orbit are a type of intraosseous meningiomas.\(^1,2,3\) Though the CT features are characteristic of the tumor, a preoperative diagnosis may be difficult. So far only five cases of intradiploic orbital roof meningioma have been reported in the literature.\(^3\)

**Case Report**

A 12-year-old male child presented with moderate proptosis of the left eye for four months. There was no history of preceding trauma. The neurological examination revealed no abnormality. The proptosed left eyeball was pushed infero-medially. There was no bruit or thrill over the eyeball. Plain radiographs of the skull showed a homogenous opacity of the orbital roof (Figure 1). The CT scan showed a hyperdense, non-enhancing tumor involving the left orbital roof (Figure 2). Lateral reconstruction images confirmed the presence of orbital roof tumor (Figure 3).

The lesion was approached by a basal, extradural, subfrontal craniotomy. The two tables of the orbital roof were expanded and removing the outer table exposed the tumor. The tumor was capsulated and firm in consistency and was typical of a fibroblastic meningioma. A complete excision of the tumor was done with minimal bleed-

Figure 1: Antero-posterior view of the plain X-ray skull showing plum-like opacity involving the orbital roof.

Figure 2: Axial contrast computerized tomography scan showing hyperdense, non-enhancing lesion involving the left orbital roof.
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ing. The inner table of the orbital roof was eroded by the tumor; however, the periorbita was normal. The bony lamina was removed up to the supraorbital rim to allow for complete reduction of the exophthalmos. Posteriorly, the optic canal was opened for a possible intracanalicular component of the tumor. A part of the tumor was then removed from the ethmoidal sinus. However, the tumor was distinctly separate from the ethmoidal mucosa. The open ethmoidal cavity was filled with bone powder. The inner table of the split calvarial frontal free bone flap was used to reconstruct the orbital roof and the supraorbital rim (Figure 4). The outer table was replaced to cover the vault. The post-operative course was uneventful. There was no evidence of any tumor on the follow-up plain X-rays. Histopathological examination revealed psammomatous meningioma (Figure 5). There was evidence of tumor infiltration into the bone but the periorbita was normal. At 1 year follow-up the left eye exophthalmos had regressed completely. There was no recurrence of tumor.

Discussion

Meningiomas originating from locations other than the meninges are termed as ectopic, extracalvarial or epidural meningiomas. Winkler, in 1904, first described a meningioma originating in an extradural location. Ectopic meningiomas although rare are found in the neck, the orbit, the paranasal sinuses, the nasal cavities and the cranial bones. Intraosseous meningiomas commonly arise from the diploe of the calvarium and exhibit osteoblastic activity. Hence, they manifest as hyperostotic lesions. They usually occur at the suture line. The exact pathogenesis of the ectopic meningiomas is unclear. It is postulated that they arise from the arachnoidal cell nests captured in the extradural tissues during the embryonal development or after cranial trauma with fracture and dural tear. Although intraosseous meningiomas commonly involve the calvarial bone, they rarely occur in the orbital roof and sphenoid bone. Intradiploic orbital roof meningiomas are extremely rare tumors and so far only 5 cases have been reported in the literature. All cases, including our case, presented with only exophthalmos on the side of the tumor. The common age of presentation was in the early second decade of life. They all presented as a plum-like opacity of the orbital roof on plain X-rays. Coronal and lateral CT images are necessary to clearly demonstrate the intradiploic involvement of the orbital roof. This observation may be otherwise missed on axial images.

The intradiploic orbital roof meningioma differs from the intraorbital meningioma in clinical presentation and radiological features. Besides proptosis, visual loss is also a common presenting feature in intraorbital meningioma. The common differential diagnosis would include fibrous dysplasia and osteoma. The histological picture in all the 6 cases reported so far, including our case, revealed Psammomatous meningioma with invasion of the bone by the meningiomatous tissue. During surgery, intradural exploration should be done if there is dural involvement on gross examination. The ethmoidal sinus mucosa was not involved by the tumor in any of the reported cases. To cure the exophthalmos it is necessary to excise the entire orbital roof involved by the tumor. The surgical exposure is improved by removing the orbital rim. Reconstruction of the orbital roof by an autogenous split calvarial bone graft harvested during surgery is necessary to prevent occurr-
rence of the pulsatile exophthalmos and for good cosmetic results.\textsuperscript{3} The other alternative methods to reconstruct the orbital roof are frontalis myofacial flap and wire mesh screen.\textsuperscript{3,11}

In conclusion, intradiploic orbital roof meningiomas, although extremely rare, have a good prognosis when excised completely. Reconstruction of the orbital roof is mandatory to prevent pulsatile exophthalmos and for better cosmetic results.

References


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