Case Report

Diploic mature teratoma originating from the orbital roof: An extremely rare case report

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Abstract

Background: Orbital mature teratoma is a rare congenital tumor.

Case Description: A 37-year-old woman presented with ophthalmalgia predated by years of progressive incongruous right eye position, diplopia, and restricted extraocular movement. Neuroimages revealed a right orbital mass originating from the orbital roof. After resection, histopathology revealed the mature teratoma.

Conclusion: To the authors’ best knowledge, this is the first documented diploic origin of mature teratoma at the orbital roof.

Key Words: Diploic origin, orbit, teratoma

INTRODUCTION

Craniofacial teratomas are rare tumors and they are believed to be congenital lesions due to the abnormal distribution of germ cells during the 3rd to 5th weeks of gestation.1 Occasionly, they could be slow growing leading to presentation in the adulthood.7 Orbital teratomas account for 1% of all orbital tumors in childhood and are usually localized within orbit and without bony involvement.3,5 We present a very rare case of diploic mature teratoma originating from the orbital roof in a young woman.

CASE HISTORY

A 37-year-old woman presented with a history of right eye pain of few days duration. There was, however, a background history of incongruous position of her eyes and diplopia of over 10 years duration. The protrusion was noted to have worsened weeks leading to presentation to a hospital where cranial computed tomography (CT) and magnetic resonance imaging (MRI) showed a right orbital mass causing exophthalmos. The mass was partly enhanced and it was predominantly intraosseous located at the orbital roof [Figures 1 and 2]. She was subsequently referred to our service. On presentation, she had lagophthalmos. Her visual acuity was 20/16 in the right eye and 20/20 in the left eye. No visual field defect was detected. She had diplopia at upward and downward gaze only. She subsequently underwent a surgery through the right frontotemporal craniotomy and superior orbitotomy. The tumor originated from the intraosseous region but the periorbit was intact. The tumor was completely removed. Postoperatively, the exophthalmos, eye pain, lagophthalmos, and diplopia
improved. The histopathologic diagnosis was compatible with mature teratoma [Figure 3]. There has been no recurrence for 6 months after surgery.

DISCUSSION

Here, we report a rare case of mature teratoma that originated from the diploic orbital roof. Orbital teratomas are rare, usually presenting with unilateral proptosis of eyeball in neonates.\(^3\) They are rapidly progressive and induce secondary damage to eyeball due to mass effect.\(^3\) A slight predominance of the left orbit was reported and the female sex was also predominantly affected.\(^2\) Histologically, orbital teratomas are usually benign with well differentiated tissues representing two or three germinal layers.\(^2,3,5\)

In general, orbital mature teratomas are purely orbital location without intracranial involvement.\(^2\) The intracranial extension is rarely reported.\(^3,5\) In addition, mature teratoma rarely develops at the calvarium.\(^4\) Therefore, diploic mature teratoma originating from the orbital roof is an extremely rare entity. To our knowledge, this is the first report of diploic mature teratoma originating from the orbital roof. We speculate that the abnormal migration of primordial germ cells into diploic tissue of the orbital roof might be responsible for the occurrence of diploic mature teratoma.

The mature teratoma usually consists of solid tissues with encapsulated cyst. Radiologic features of mature teratoma are nonspecific because they depend on containing different well-differentiated components such as soft tissue, cartilage, or bone. Therefore, mature teratomas have mixed density and intensity on computed tomography and magnetic resonance imaging, respectively, whereas contrast enhancement is variable.\(^7\) In addition, calcification is found in half of the reported cases of mature teratoma. In the present case, neither calcification nor encapsulated cyst were found on CT and MRI. The only peripheral enhancement was seen on MRI.

Complete resection for mature teratoma leads to a surgical cure and prognosis is good.\(^6,7\) If teratoma contains immature tissues, chemotherapy, and radiotherapy are recommended.

CONCLUSION

An extremely rare case of diploic mature teratoma arising from the orbital roof was presented.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.
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Conflicts of interest
There are no conflicts of interest.

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