Glial heterotopia in an adult: A rare orbital mass

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Heterotopic glial tissue is very rare in the orbit. Our case was an adult, which is unique since most cases reported in literature involve children. We describe a case of a 60-year-old man who presented with an orbital mass, which histopathologically revealed heterotopic glial tissue.

Key words: Ectopic brain tissue, glial heterotopia, orbit

Ectopic brain tissue being found in the orbit is very rare. It usually presents along the mid-line structures, especially nasal.[1] Few cases of ectopic glial tissue reported in literature mainly involved infants or children. Our case is unique that it was found in an adult.

Case Report

A 60-year-old man presented to our institute with a swelling in the superior medial aspect of his right orbit. It has been noticed for the last 1 year and has been gradually increasing in size. He had associated complaints of double vision. There was no history of defective vision, pain, or sudden increase in size. On examination, the best-corrected visual acuity was 20/20 in both eyes. A soft to firm globular mass in the superomedial aspect of the orbit displaced the right eye downward and outward. It was not compressible or reducible, and the deeper margins were not palpable [Fig. 1]. Anterior segment and posterior segment of both the eyes were normal. Extraocular movements in the right eye showed restriction in elevation, levo-elevation, and adduction.

On magnetic resonance imaging, a 39 mm × 26 mm lobulated well-encapsulated lesion was seen in the superomedial aspect of the right orbit. It was hyperintense in T2 and flair sequence and isointense in T1 imaging [Fig. 2]. The mass displaced the medial rectus, encircling the superior rectus, and was extra- and intra-conal. An orbitotomy was performed after obtaining consent the mass was excised.

Histopathological examination revealed areas of hemorrhage and clusters of short spindle cells in eosinophilic fibrillary background [Fig. 3]. Cells have elongated nucleus and scanty eosinophilic cytoplasm along with foci of myxoid stroma and irregular vascular channels, thick- and thin-walled lined by flat endothelial cells. A histopathological diagnosis of glial heterotopia was made.

Postoperatively, the patient had improved extraocular motility, no diplopia, and the proptosis showed regression. At 1 year follow-up, the patient showed no sign of any recurrence.

Discussion

Glial heterotopia is abnormally located collection of normal glial tissue distant to the central nervous system. The ectopic glial tissue is discontiguous with the cranial cavity.[2] It most frequently occurs in the nasal midline. The most common location is around the nasal cavity, and it can occur in sites such as ethmoid sinus, middle ear, pharyngeal area, parapharyngeal space, pterygopalatine fossa, submandibular region, scalp, head, neck, and lung. Glial tissue in the orbit is very rare.[3]

They are generally present at birth but can manifest at any time in life. Of the previously reported orbital glial heterotopia, the age of presentation has varied between 15 days and 20 years.[4] Our patient presented in the sixth decade, and the glial tissue was present in the orbit without any bony defect. There is only one other case reported in
literature in a 59-year-old man who presented with optic disc swelling, and the orbital mass was found to be ectopic glial tissue.[1]

The mass is a benign slow-growing choristoma. Patients may present with varying symptoms of proptosis, diplopia, ptosis, or lid swelling due to mass effect.[1,5] Our patient presented with a slow-growing orbital mass causing dystopia and diplopia.

The most widely accepted theory in the pathogenesis of heterotopic glial tissue is that there is embryological herniation of glial tissue through a bony defect which subsequently may close, leaving no communication with the cranial cavity.[1,5] They are also known as glial choristomas, neurogenic hamartomes, or ectopic gliomas. On histopathology, heterotopic glial tissue found in the orbit is composed of astrocytes, gemistocytic astrocytes, fibrovascular connective tissue, lacrimal gland, cerebellar tissue, primitive retina, and skeletal muscle in varying proportions. There can be associated laminated calcific bodies or calcospherules; it can be cystic filled with cerebrospinal fluid or clear fluid. Histopathology in our case revealed the presence of myxoid tissue along with glial tissue. The muscle tissue is believed to be a choristomatous component of the tumor. Some authors opine that this could be dysembryogenic in origin, and some opine that it could have got admixed with the developing muscle.[4,6]

These masses have local growth potential and no known malignant potential. The accepted role of management is maximum possible excision. Surgery may be difficult and bony defects have to be looked for, which may then need multi-speciality approach. Surgeons should be aware of such orbital masses, presenting later on in life.[6]

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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