Chondroid Syringoma Of The Orbit -A Peculiar Tumor Imaging And Its Management

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ABSTRACT

Chondroid syringoma (CS) of the orbit is virtually a rare benign neoplasm. As far as the latest information is concerned with updated knowledge and visualizing techniques this is the rare case being reported in the existing British literature . The examining authors now bring to light a case of a man aging 60 years affected with orbital Chondroid syringoma. This cited tumor grew at a low speed within an interval of over 8 years and thus giving rise to indolor with exopthalmos of the right eye. Computed tomography revealed a well circumscribed heterogeneously enhancing solid mass lesion at supero-lateral aspect of right orbit with no bony erosion, calcification and periosteal reaction . On the eve of surgery the developed mass was definitely circumscribed, very rigid, extraconal and not invading or adhering to nearby organs. The tumor was completely removed from all sides. No recurrence was ever detected even after adopting two year's follow up. An unusual case of a patient is being hereby reported who underwent resection of an intraorbital chondroid syringoma completely. Therefore it becomes mandatory to very well include chondroid syringoma in the differential diagnosis of intraorbital tumors. The common sites of their prevalence are the cheek, scalp, chin, upper lip, nose and the forehead. Chondroid syringoma has been found to be affecting middle aged or older male patients. Resection in all respects remains the best therapeutic option to get rid of any kind of recurrence and close follow up can be ascertained due to rarely seen malignant transformation was potency may be still feasible.

Keywords : *chondroid syringoma, intraorbital , benign , tumor , Computed tomography*

INTRODUCTION

The differential diagnosis of chondroid syringoma can be achieved through peeping in to other benign orbital lesions including lipoma , pleomorphic adenoma , neurofibromas, histocytic tumors, desmoids, meningioma and cavernous hemangioma. The knowledge derived from magnetic resonance and depending upon our corroborative skill of magnetic resonance imaging. The tumor was extremely isointense on T1 weighted imaging, hyperintense on T2 weighted imaging with extraconal mass lesion in right orbit displacing the optic nerve medially and no evidence of diffusion restriction. The globe was displaced anteromedially . Chondroid syringoma has been considered rare mixed tumors of sweat glands which were detected and established for the first time by Billroth in 1859 being either benign or malignant. Histological findings such as cytological atypia , satellite tumor nodules , tumor necrosis, infilterative margins and involvement of deep structures have been considered distinct characterstics of malignant transformation. The diagnosis was also correlated to histopathological examination, the lesion was apparently nodular as well as differentiation was equally nodular. Moreover differentiation was duly observed towards the adnexal ductal epithelium with chondromyxoid and adipocytes segregation in the stroma. The term "chondroid syringoma" was initially coined by Hirsh and Helwig in 1961 to explicitly explain a specific type of skin tumor arising apparently in the apocrine sweat glands especially comprising of glandular elements and stroma very much similar to cartilage. Incidence of chondroid syringoma has been very well reported in less than 0.01% of primary tumors of the skin. Clinically expressed chondroid syringoma presents as a typical slow growing, painless, rigid, non ulcerated subcutaneous or intracutaneous nodule.[3,4,5]

The lesion generally measures 0.5-3 cm in diameter. However, even larger forms chondroid myxoma have been very well depicted. Occasionally these malignant types have been generally seen in young females bearing a predilection to occur especially on the trunk or extremities. Intensive care

and steady follow up of these tumors is the need of the hour due to high risk of malignancy and recurrence. Oftenly these tumors have a bulky mass measuring 3 cm or above and tend to invade locally.[1,2]

Hirsch and Helwig applied these appellation of chondroid syringoma in 1961 as a mark of presence of varied constituents of sweat glands found to be arranged in cartilaginous stroma. Chondroid syringoma of the orbit has been rarely observed. On minute viewing partial visual field defect very well existed. Cranial CT scan revealed a well circumscribed heterogeneously enhancing superolateral solid mass of right orbit with no bone infiltration, calcification, erosion and periosteal reaction. The diagnosis of chondroid syringoma got confirmed following application of histopathological test. Preoperative diagnosis further established pleomorphic adenoma and adenoid cystic carcinoma. No hemorrhage or necrosis were noticed in the adjoining organs. The optic nerve had been displaced medially by the intervening mass. The globe was displaced anteromedially . Histologically the lesion was nodular with concrete differentiation towards the adnexal ductal epithelium associated with chondromyxoid and adipocyte differentiation in the stroma . Kitazawa T et al have already reported the first case of a woman with peculiar complaint of left exopthalmos and diplopia in 1999. A small rigid mass was found to be palpable in the left lateral canthal region. CT and MRI jointly revealed an extraconal mass extending to the malar subcutis and in association with compressed globe. The tumor was resected conveniently and diagnosis of benign chondroid syringoma had been further confirmed by applying histopathological tests. No palpable mass had been detected in this cited case. [3,4,5]

RESULTS

One man aging 60 years presented with a history of right exopthalmos consistently for 8 years and deterioration of visual acuity in all respects in the past 8 months.(FIGURE 1) Moreover, medical and ocular histories of the patients had been astonishable, unbelievable and unremarkable. Physical examination revealed no abnormality in the prescribed system other than ocular disorders. An indolor right exopthalmos was noticed without downward displacement of the globe. Abduction of the right eye was confined to a little extent.

- swelling of right eye for last 8 years gradually increasing in size .
- progressive diminution of vision for last 8 years.
- watering and redness of eye for 2 months.
- On examination-firm, non tender exophytic mass arising from orbital tissue fixed to underlying structure.

FIGURE 1

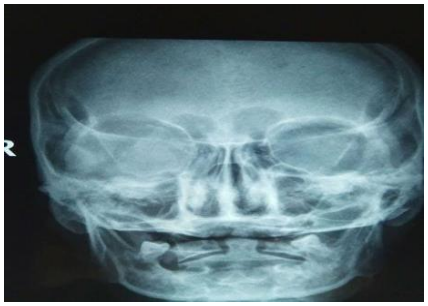
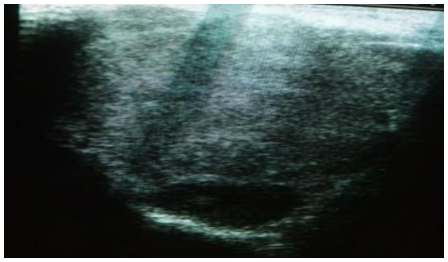
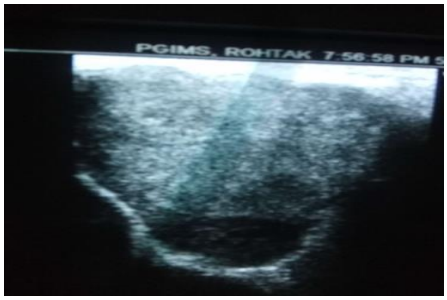


FIGURE 2 : X ray orbit (AP view)-

- Soft tissue density in right lateral aspect of orbit with no bony involvement/reaction.
- Widening of lateral margin of orbit.
- No periosteal reaction.

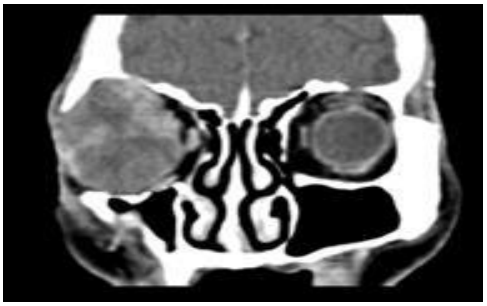


USG IMAGE: A.



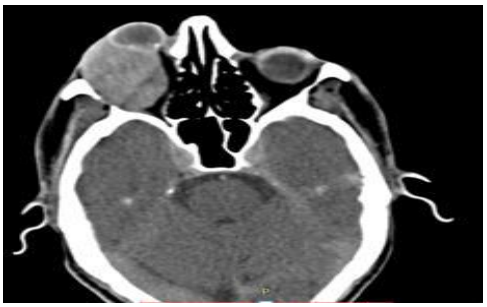
USG IMAGE: B.

FIGURE 3 : USG Orbit (A-B) – An extraconal mass lesion in right supero-lateral orbit displacing eye ball anteromedially and showing no flow on color Doppler without any surrounding bony erosion or globe infiltration.

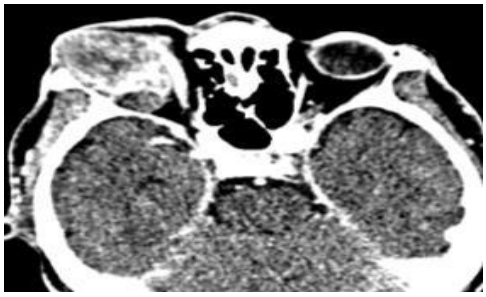


CT IMAGE : A.

CT IMAGE : B.

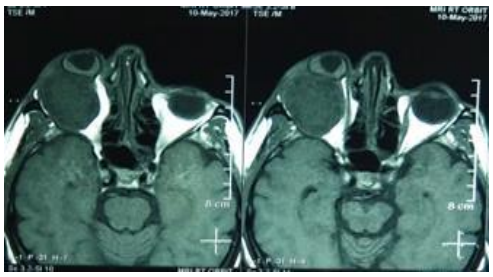


CT IMAGE : C.

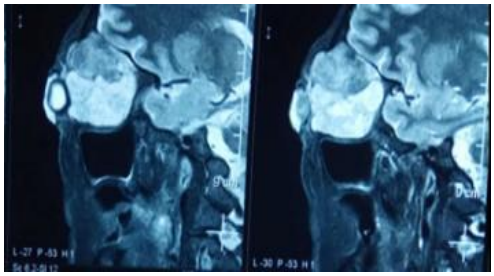


CT IMAGE : D.

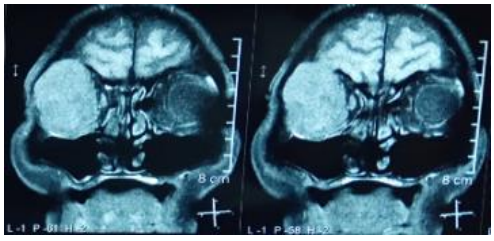
FIGURE 4 : CT Orbit (A- D) CHONDROID SYRINGOMA - A well circumscribed heterogeneously enhancing solid mass lesion at supero-lateral aspect of right orbit. No bony erosion. No calcification and periosteal reaction . The optic nerve had been displaced medially by the intervening mass. The globe was displaced anteromedially .



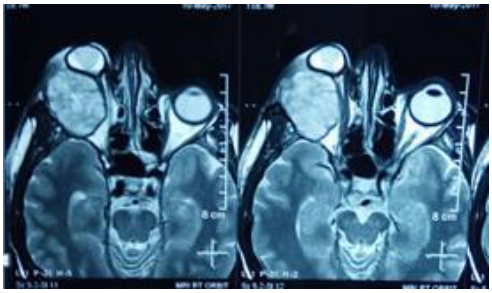
MRI IMAGE : A.



MRI IMAGE: B.



MRI IMAGE : C.



MRI IMAGE : D.

FIGURE 5 : MRI Orbit(A-D) CHONDROID SYRINGOMA –T1 isointense , T2 hyperintense extraconal mass lesion in right orbit displacing optic nerve medially . No diffusion restriction.

DISCUSSION AND CONCLUSION

Chondroid syringoma is the rare of the rarest benign skin appendage tumor mentioned and explained for the first time by Billroth in 1859 for a group of aggregated leading tumors of the salivary glands containing varying volumes of mucoid and also varied amount of cartilaginous mass. An unusual case of a patient is being hereby reported who underwent resection of an intraorbital chondroid syringoma completely. The histological findings, clinical presentation followed by accurate management has already been discussed by taking into consideration latest available review material. MRI revealed an extraconal round mass lesion in right orbit. On T1 weighted imaging isointensity had been actually viewed corresponding to hyperintense signal on T2 weighted imaging with displacement of optic nerve medially.The eyeball appeared to be anteromedially displaced. The tumor was removed in toto through lateral orbitotomy with drastically reduced orbital rim osteotomy. Virchow and Minssen have already mentioned these as mixed tumors having both epithelial and mesenchymal origin. Therefore, complete excision of the cited tumor is an urgent need of the hour for its weeding out, termination, culmination to have a strong vigil consistently towards its long term follow up to provide sooth and comfort to the afflicted patient who has already suffered a lot.[1,2]

Initial postoperative CT image depicts overall removal with exemplary orbital reconstruction. The postoperative course was certainly casual. The patient was discharged 7 days following normal operation devoid of any complication and intrication. Eye motility,visual acuity and cosmetic appearance had been satisfactorily improved. No recurrence had been visualized following two year of follow up with regards to magnificent treatment. On the eve of surgery the mass was extraconal, very rigid followed by normal enucleation and with no invasion or adhesion to other pertinent adjoining structures including orbital bone , globe, extraocular muscles, skin and lacrimal gland.

The exact sites of predilection pertaining to chondroid syringoma are on the head and neck region especially nose, cheek or skin above the lip. To a lesser extent this tumor can grow on the scalp, hands , axillary regions, eyelids, forehead, feet, abdomen, vulva, penis and scrotum. Moreover, its orbital location has also been visualized in exceptional cases. Therefore, a medline data base had been under the question while focusing on the key words chondroid syringoma orbit only two cases have been still established . Henceforth the treatment of choice is to sort out the remedy and to get rid of this unique problem or for its elimination or culmination performance lies in adopting and reviving surgical techniques with complete excision without aesthetic or functional disruption of adjoining structures. We focused our attention on this particular patient to pin point that this tumor can be regarded as a rare possibility in the differential diagnosis related to orbital tumors. Chondroid syringoma could be asserted to closely review differential diagnosis relating to intraorbital tumors especially in the individuals of middle age suspected to be suffering from this peculiar disorder. Radiological features of chondroid syringoma have not been considered equally suggestive corresponding to the histological findings. But the MRI features in particular have been found to be non specific, hence depict the anatomic extent with more accuracy such as identification of tissue origin more clearly , depth of invasion in relation to living structures in close viscinity especially muscles and bones. Generally chondroid syringoma lesions may not establish clinically or found to be radiological distinctive as the diagnosis has been equally based on histological studies. [2,3,4]

Lateral orbitotomy has been considered a safe practice in the surgical management of lesions especially in the lateral orbital region and if the lesions are extraconal in particular. It has been further established in incisional biopsy that chondroid syringoma is a type of benign tumor. However very rare cases of malignant chondroid syringoma have

been reported. As these malignant types of tumor recur occasionally.

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No words can ever express my deep sense of gratitude for my parents & my younger brother, for their affections, endurance, inspiration, support, unending blessings, innumerable sacrifices and unceasing encouragement that has moulded me into the person i am today. the expression of my gratitude's will remain incomplete if i fail to register my deep sense of indebtness to my family members without whose perseverance and affection it would have not been possible for me to undertake this ardous assignment.

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