



BENIGN CHONDROID SYRINGOMA OF THE ORBIT LEADING TO EXOPHTHALMOS: AN EXTREMELY RARE ENTITY

Urmil Chawla¹, Jyoti Deswal*², Ashok Kumar Khurana³, Nisha Marwah⁴, Jyotsna Sen⁵, Hemlata T Kamra⁶

¹Professor, Regional Institute of Ophthalmology, PGIMS, Rohtak, Haryana, India.

²Senior Resident, Regional Institute of Ophthalmology, PGIMS, Rohtak, Haryana, India.

³Senior Professor, Regional Institute of Ophthalmology, PGIMS, Rohtak, Haryana, India.

⁴Professor, Department of Pathology, PGIMS, Rohtak, Haryana, India.

⁵Senior Professor, Department of Radiology, PGIMS, Rohtak, Haryana, India.

⁶Professor, Department of Pathology, KalpanaChawla Government Medical College and Hospital, Karnal, Haryana, India.

ABSTRACT

Chondroid syringoma is a rare benign neoplasm and its occurrence in orbit is extremely rare. We report a case of 60 year old Indian male who presented with complete loss of vision with gradual progressive protrusion of right eyeball for the past 6 months. The patient also had firm round swelling over temporal aspect of right brow for the past 6 years. Radiological imaging showed a well defined (48x32mm) lobulated mass lesion of heterogenous signal intensity in right retrobulbar area which causes expansion and erosion of lateral orbital wall and proptosis of right globe. Right lateral rectus could not be identified separately from the mass. Total excision of the mass was done with enucleation of the atrophic globe. Histopathological examination revealed it to be Chondroid syringoma. Complete resection of the tumour is the best treatment option to prevent recurrence and close follow-up is recommended as malignant transformation is possible.

Key words: Chondroid syringoma, Polymorphic adenoma, Enucleation.

Access this article online

Home page:

<http://www.mcmed.us/journal/ijacr>

DOI:

<http://dx.doi.org/10.21276/ijacr.2018.5.1.3>

Quick Response code



Received:25.01.18

Revised:12.02.18

Accepted:15.02.18

INTRODUCTION

Chondroid syringoma (CS) is a rare tumour of skin arising from sweat glands which was first described by Billroth in 1859. It is a mixed tumour, also called pleomorphic adenoma of skin, having both benign and malignant forms. The term Chondroid syringoma was coined by Hirsch and Helwig in 1961 because of the presence of sweat gland elements which are set in a

cartilaginous stroma [1]. The commonest sites for presentation are scalp, cheek, nose, upper lip, chin and the forehead. Orbital Chondroid syringoma is extremely rare. Here, we report an unusual case who underwent complete resection of intraorbital CS. To the best of our knowledge, our case is the third case reported in the English literature.

Case Report

A 60 year- old Indian male presented with gradual progressive protrusion of right eyeball for the past 6 months along with gradual complete loss of vision in right eye. The patient also had round, painless swelling over temporal aspect of right brow for the past 6 years. The

Corresponding Author

Jyoti Deswal

Senior Resident, Regional Institute of Ophthalmology, PGIMS, Rohtak, Haryana, India.

Email: jyoti_deswal@yahoo.co.in

protrusion of eye was associated with mild boring pain, redness and occasional watering from the eye. There is no history of any trauma, ocular or head surgery in the past. The patient is a chronic smoker, non- alcoholic, vegetarian by diet and has no systemic ailment.

On clinical examination, patient was well-nourished and moderately built with stable vitals. Systemic examination was within normal limits. On ocular examination, there was no perception of light in right eye. The whole eyeball was dystrophic and was axially proptosed. The anterior surface including cornea was keratinized (Figure-1). Ocular movements were restricted in all gazes. Over superotemporal margin of orbit, there was a round, firm, non-tender mass of size approx 2.5 x 1.5 cm, which was immobile, adherent to underlying structures, non-compressible and non-fluctuant. The fellow eye had best corrected visual acuity of 6/18 due to immature senile cataract, rest of the examination was normal.

Radiological imaging i.e CT scan showed a well defined (48x32mm) lobulated mass lesion of heterogenous signal intensity in right retrobulbar area causing expansion of lateral orbital wall and proptosis of right globe. Right lateral rectus could not be identified separately from the mass. Anteriorly, the mass was extending into pre-septal and pre-maxillary space. The globe appeared small and atrophic with wall calcification (pthisis) (Figure-2A). MRI showed a round mass that was extraconal, showing hypointensity on T1-weighted imaging, slightly

hyperintensity on T2-weighted imaging and enhanced after intravenous contrast administration. The globe was displaced anteriorly. The optic nerve was medially displaced and compressed by the mass. Contralateral orbit appeared normal. (Figure-2B, 2C and 2D). Our preoperative differential diagnoses was pleomorphic adenoma and adenoid cystic carcinoma

Enucleation of the atrophic eyeball with removal of the tumor was done under general anaesthesia. The superotemporal mass at eyebrow appeared attached to the orbital mass and was also removed. The excised masses varied from 1.5cm to 4 cm in diameter and was extraconal, well encapsulated, firm in consistency, grayish-white in colour. (Figure-3). It showed no invasion or adhesion to other structures, including skin, globe, extraocular muscles, lacrimal gland, and orbital bone. Gross pathologic analysis revealed well-encapsulated tan tissue with no areas of hemorrhage or necrosis noted. Histologically, the lesion was nodular, and there was differentiation toward the adnexal ductal epithelium with chondromyxoid and adipocytic differentiation in the stroma. Histopathological examination confirmed it to be benign mixed tumour suggestive of Chondroid syringoma. (Fig 4). Early post-operative CT image showed complete removal of the tumor. Post-operative period was uneventful. Artificial eye shell was applied after 2 weeks of surgery for cosmetic rehabilitation. There is no evidence of recurrence after one year of follow-up.

Fig 1. Patient’s photograph showing a mass over superotemporal aspect of right eyebrow and axially proptosed dystrophic globe.



Fig 2A. CT orbit axial view showing a well-defined (48x32mm) lobulated mass lesion of heterogenous signal intensity in right retrobulbar area, 2B, 2C, 2D-MRI images showed a round mass that was extraconal, showing hypointensity on T1-weighted imaging, slightly hyperintensity on T2-weighted imaging and enhanced after intravenous contrast administration.

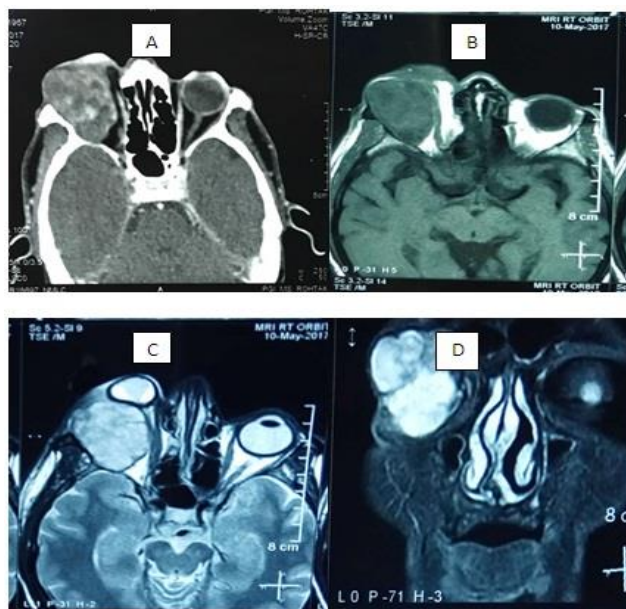
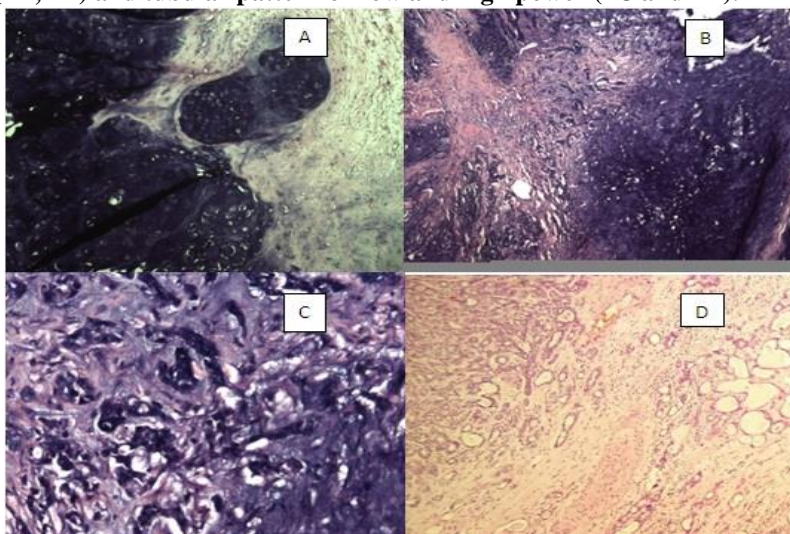


Fig 3. Photograph of surgically excised mass, well encapsulated, firm in consistency, grayish-white in colour.



Fig 4. Photomicrograph (Hematoxylin and Eosin stain) of surgically excised specimen showing chondromyxoid stroma on low and high power (4A, 4B) and tubular pattern on low and high power (4C and 4D).



DISCUSSION

Chondroid syringoma is a benign tumour of skin appendages. It was first described by Billroth in 1859 for a group of tumors of the salivary gland that contained varying amounts of mucoid and cartilaginous material [2]. Chondroid syringoma usually affects middle-aged and older people and there is slight male preponderance. It is very rare as its incidence has been reported as less than 0.01% of primary tumors of the skin [3].

Chondroid syringoma has silent clinical presentation as slow growing, solitary, painless, firm mass or papule of size varying from 0.5 to 3 cm localized in dermis or subdermis. The surface is non-ulcerated and can be erythematous, purple or skin coloured with its base free from underlying tissues [4]. The head and neck region is most commonly involved, particularly cheek, nose and upper lip skin. Other sites where occurrence of CS has been reported are scalp, hand, foot, eyelid, forehead, axilla, abdomen, penis, vulva and scrotum [5-7]. Orbital involvement is extremely rare. To the best of our knowledge only two cases have been reported so far. First case was reported in 1999 by Kitazawa et al in a young female who presented with left eye exophthalmos. The

mass was firm in consistency, extra-conal in position and was palpable in left canthal area. The tumour was excised successfully and histopathology confirmed the diagnosis of Chondroid syringoma [8]. Belfquih et al reported the second case of Chondroid syringoma in 2012 in a 41 year-old female with a firm well circumscribed mass located extraconally in superolateral orbit. The mass was excised by lateral orbitotomy. Histopathological examination revealed it to be Chondroid syringoma. No recurrence of tumour was seen with one year of follow up [9]. Our case is the third case of orbital Chondroid syringoma reported so far in English literature.

Histopathologically, it is a mixed tumour consisting of epithelial and mesenchymal elements arranged in tubuloalveolar and gland like structures with two or more cuboidal cell lines, islands of cuboidal or polygonal cells in a fibroadipoid, chondroid, hyaline or mucinous hypocellular stroma [10]. Headington described two histological variants of the tumour - Apocrine and Eccrine [11]. The apocrine type is more common and is characterized by haphazardly arranged broad sheets of squamous epithelial cells or tubular branching lumina lined by two layers of epithelial cells. The eccrine type is

characterized by homogeneously arranged epithelial components with small tubular lumina lined by a single layer of epithelial cells. Immunohistochemistry shows focal positivity for keratin, vimentin, desmin and S-100 protein [12]. Hirsch and Helwig also proposed five histopathological criteria for its diagnosis: 1) nests of cuboidal or polygonal cells; 2) intercommunicating tubuloalveolar structures lined with two or more rows of cuboidal cells; 3) ductal structures composed of one or two rows of cuboidal cells; 4) occasional keratinous cysts; 5) a matrix of varying composition. Chondroid syringomas may have all five characteristics or manifest only some [1]. Four criterias were found in our case. Malignant transformation is rare and characterized by nuclear atypia, increased mitotic figures, infiltrative margins, satellite tumour nodules and tumour necrosis [13]. Tumour sizes more than 3cm have a greater likelihood of malignancy [14].

Radiological features are non-specific but are helpful in depicting anatomical extent of the growth, invasion and relation to adjacent structures. Chondroid syringoma may be confused with other benign tumours of epidermal or mesenchymal appendages such as pleomorphic adenoma, dermoid, neurofibroma, lymphangioma, cavernous hemangioma, lipoma, sebaceous cyst, lymphoma and histiocytic tumours [15]. Histopathological examination of the excised tumour remains the mainstay for confirmation of the diagnosis.

Various treatment modalities have been proposed for Chondroid syringoma like electrodesiccation and

vapourisation with CO₂ or Argon lasers but complete surgical resection of the tumour, as done in our case, is the treatment of choice [16, 17]. This should be followed by regular follow-up of the patient to look for recurrence and malignant transformation, though rare, but possible [18, 19].

CONCLUSION

Chondroid syringoma is a rare tumour of skin and its appendages. It should be kept in the list of differential diagnosis of intra-orbital tumours especially in middle - aged males. For, such a lesion, excisional biopsy without destroying esthetic and functional structures is the preferred diagnostic as well as therapeutic approach. Wide excision and long term follow-up is the mainstay of its management.

STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors

ACKNOWLEDGEMENT

Nil

CONFLICT OF INTEREST

No interest

REFERENCES

- Hirsch P and Helwig EB. (1961). Chondroid syringoma: mixed tumour of the skin, salivary gland type. *Arch Dermatol*, 84, 835-847.
- Sungur N, Uysal A, Gümüs M, Kocer U. (2003). An unusual Chondroid syringoma. *Dermatol Surg*, 29, 977-979.
- Yavuzer R, Basterzi Y, Sari A, Bir F, Sezer C. (2003). Chondroid syringoma: A diagnosis more frequent than expected. *Dermatol Surg*, 29, 179-181.
- Bekerecioglu M, Tercan M, Karakok M, Atik B. (2002). Benign Chondroid syringoma: A confusing clinical diagnosis. *Eur J Plast Surg*, 25, 316-318.
- Poku JW, Sant GR, Ucci AA. (1996). Chondroid syringoma of the scrotum. *J Int Med Res*. 24. 482-486.
- Nemoto K, Kato N, Arino H. (2002). Chondroid syringoma of the hand. *Scand J Plast Reconstr Surg Hand Surg*, 36, 379-381.
- Takamitsu O and Shinichi W. (1997). Histogenesis of mixed tumor of the skin, apocrine type: immunohistochemical study of keratin expression. *Am J of Dermatopathol*, 19, 456-461.
- Kitazawa T, Hataya Y, Matsuo K. (1999). Chondroid syringoma of the orbit. *Ann Plast Surg*, 42, 100-102.
- Belfquih H, Mostarchid BE, Oukabli M, Akhaddar A, Boucetta M. (2012). Benign Chondroid syringoma of the orbit: a rare cause of exophthalmos. *Head & Face Medicine*, 8, 8.
- Chen AH, Moreano EH, Houston B, Funk FG. (1996). Chondroid syringoma of the head and neck: clinical management and literature review. *Ear Nose Throat J*, 75, 104-108.
- Headington JT. (1961). Mixed tumors of the skin: eccrine and apocrine types. *Arch Dermatol*, 84, 989-996.
- Ferreiro JA, Nascimento AG. (1995). Hyaline-cell rich Chondroid syringoma: "a tumor mimicking malignancy". *Am J Surg Pathol*, 19, 912-917.
- Gunduz K, Demirel S, Heper AO, Gunalp I. (2006). A rare case of atypical Chondroid syringoma of the lower eyelid and review of the literature. *Surv Ophthalmol*, 51, 280-285.
- Barnett MD, Wallack MK, Zuretti A, Mesia L, Emery RS, Berson AM. (2000). Recurrent malignant Chondroid syringoma of the foot: a case report and review of the literature. *Am J Clin Oncol*, 23, 227-232.

15. Chandran S, PillaiAM. (1990). Chondroid syringoma scalp A case report. *Indian J Cancer*, 27, 97-100.
16. Jordan DR, Nerad JA, Patrinely JR. (1989). Chondroid syringoma of the eyelid. *Can J Ophthalmol*, 24, 24-27.
17. Karam P and Benedetto AV. (1996). Syringomas: new approach to an old technique. *Int J Dermatol*, 35, 219-220.
18. Nather A and Sutherland IH. (1986). Malignant transformation of a benign cutaneous mixed tumour. *J Hand Surg*, 11, 139-143.
19. Toback TG and Toback CJ. (1989). Chondroid syringoma: A review of the literature and case report. *J Am Podiatr Med Assoc*, 79, 563-565.

Cite this article:

Urmil Chawla, Jyoti Deswal, Ashok Kumar Khurana, Nisha Marwah, Jyotsna Sen, Hemlata T Kamra. Benign Chondroid Syringoma of the Orbit Leading to Exophthalmos: An Extremely Rare Entity. *International Journal of Advances in Case Reports*, 5(1), 2018, 8-12. DOI: <http://dx.doi.org/10.21276/ijacr.2018.5.1.3>



Attribution-Non Commercial-No Derivatives 4.0 International