CASE REPORT

Lacrimal gland pleomorphic adenoma: an inconceivable diagnosis in a child

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SUMMARY

Pleomorphic adenoma is the most common epithelial tumour of the lacrimal gland in adults. It is rare in children less than 10 years. The low incidence in children can lead to low index of suspicion and increased likelihood of misdiagnosis. Incomplete treatment can lead to malignant transformation or recurrence. High index of suspicion is necessary for prompt diagnosis in children younger than 10 years. We describe a rare case of lacrimal gland pleomorphic adenoma (LGPA) in a 7-year-old child. Only four cases have so far been reported in the literature in children younger than 10 years. We emphasise the importance of CT scan and ultrasound-guided fine needle in the diagnosis of LGPA in children.

BACKGROUND

Pleomorphic adenoma is the most common epithelial tumour of the lacrimal gland.^{1 2} Lacrimal gland pleomorphic adenoma (LGPA) occurs commonly in second to fifth decades.^{3–7} LGPA is a rare tumour in children less than 10 years.⁸ LGPA constitutes 3–5% of all orbital tumours, 25% of all lacrimal gland lesions and 50% of epithelial lacrimal gland tumours.⁸

We describe a rare case of LGPA in a 7-year-old child. This case report emphasises the importance of CT scan and ultrasound-guided fine needle aspiration cytology (FNAC) in precise localisation and characterisation of superotemporal mass lesions. FNAC helps rule out pathologies like lymphoma, pseudotumour and malignancy, and thus helps formulate the appropriate treatment plan. Early diagnosis and treatment helps preserve vision and prevent future recurrences and malignant transformation.

CASE PRESENTATION

A 7-year-old boy presented with proptosis of the left eye for the duration of 2 months. The proptosis was painless, progressive and non-pulsatile. No postural variation in the proptosis was observed. There was no history of diminution of vision in the left eye. There was no history of any constitutional symptoms or trauma. Examination revealed a visual acuity of 20/20 on the Snellen chart in both the eyes. Anterior segment examination including the pupillary responses was within normal limits. The proptosis measured using the Hertel exophthalmometer showed readings of 22 mm for the left eye and 19 mm for the right eye. The left eye was displaced inferiorly and medially. The ocular movement of the left eye was restricted in upgaze. Intraocular pressure and visual fields were within normal limits.

INVESTIGATIONS

A CT scan of the left orbit demonstrated 2.7×3.1 cm sized, relatively homogenous, well-circumscribed, enhancing lesion in the extraconal compartment along the superolateral wall of the left orbit (figure 1). The differential diagnosis of lymphoma, inflammatory pseudotumour or benign tumour of the lacrimal gland was considered. Subsequently, the patient underwent ultrasound-guided FNAC of the left orbital mass (figure 2). FNAC was performed using 22-gauge needle, which revealed cellular smear mixed stromal epithelial fragments dispersed singly and in sheets. The epithelial cells showed uniform nuclear size and chromatin (figure 3). The diagnosis of the benign mixed tumour of the lacrimal gland was made and an excision biopsy was advised.

Figure 1 Coronal (A) and axial (B) contrast-enhanced CT images of the orbit revealing mildly enhancing, well-defined extraconal mass lesion in the left supero-temporal region.

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Figure 2 (A) Ultrasonography of the orbit showing a well-circumscribed, hypoechoic mass lesion without posterior acoustic shadowing confirming its solid nature. (B) Colour Doppler did not show significant internal vascularity.

DIFFERENTIAL DIAGNOSIS

Pleomorphic adenoma should be differentiated from

- ▶ Adenoid cystic tumour
- ▶ Monomorphic adenoma
- ▶ Mucoepidermoid carcinoma
- ▶ Carcinoma ex-pleomorphic adenoma

TREATMENT

En bloc excision of the mass was performed through lateral orbitotomy approach. The mass was sent for histopathology, which demonstrated a heterogeneous encapsulated tumour with the epithelial as well as mesenchymal components. No feature of malignant transformation was seen.

OUTCOME AND FOLLOW-UP

The patient did very well after the surgery with no symptoms at 1 year after the operation.

DISCUSSION

LGPA is a benign, slow-growing, epithelial tumour. It is a well-circumscribed and pseudoencapsulated superotemporal mass. It can be differentiated from a malignant lesion by the absence of associated pain and bony erosion. LGPA is commonly seen in young and middle age group. It is rarely seen in children less than 10 years.⁸ Only a few cases of LGPA have been reported in the literature in children less than 10 years of age. The differential diagnosis of an expansile lacrimal fossa masses is adenoid cystic carcinoma, lymphoma and pseudotumour. It may be difficult to differentiate pleomorphic adenoma from adenoid cystic tumour, monomorphic adenoma, mucoepidermoid carcinoma and carcinoma expleomorphic adenoma on FNAC. Orbital lobe of the lacrimal gland is involved in 90% of the cases.⁹ LGPA can rarely arise from the palpebral lobe or an ectopic lacrimal tissue.¹⁰ A modified algorithm for the management of masses of the orbital lobe of the lacrimal gland can aid in scoring and strategic planning of prudent management. The Rose and Wright modified scoring algorithm for lacrimal gland masses accords one point each in the presence of the following: duration of acute symptoms greater than 10 months, absence of persistent

pain, absence of sensory loss, a well-defined round or oval mass on imaging, absence of moulding of mass to globe or along the lateral orbital wall, absence of calcification, absence of bone invasion and presence of a small-sized tumour with long duration of symptoms. One point is deducted for each criterion that is not met. A score of +3 to +8 has been reported to favour a diagnosis of LGPA. A score of −6 to +2 supports the likelihood of a malignant-mixed lacrimal tumour, whereas a score of −8 to +2 points towards the increased probability of carcinoma.²

A CT scan aids in initial localisation and characterisation of the tumour. It can help in differentiating benign from malignant lesions, with respect to bony erosion and moulding and associated calcification.^{2 11}

Initially, a strict no biopsy technique was applied when dealing with lacrimal gland tumours as biopsy was thought to increase the risk of recurrence. Lai *et al*¹² demonstrated that FNAC differentiates between various lacrimal gland pathologies and helps in definitive management; it prevents incomplete excision of a malignant lesion and future recurrence. Verma and Kapila¹³ demonstrated FNAC to have a specificity of 98.2% and positive predictive value of 96.7%. Vagefi *et al*¹⁴ recommended a complete surgical excision of the tumour to prevent future recurrence and malignant transformation. Thus, a preoperative FNAC is prudent in diagnosing and tailoring the management of each individual case. The recurrence can further be prevented by excision of the biopsy track.

Excisional biopsy is the definitive treatment. Anterior orbitotomy is the preferred route of surgery for access to the anterior extracanal orbital space (palpebral lobe lesions). Lateral orbitotomy is an approach for deeper orbital lesions that could not be accessed through an anterior incision or require larger exposure (orbital lobe lesions). Recurrence can occur if the tumour is incompletely excised or if an incisional biopsy was performed preoperatively.^{2 11} The chances of recurrence 5 years postoperatively are 3% if completely excised with an intact capsule and 30% if incompletely removed.⁴ LGPA can serve as a locus for future development of malignant epithelial neoplasm. Around 10% of lacrimal gland tumours become malignant within 20 years of diagnosis and it increases to up to 20% after 30 years.⁹

Figure 3 Low (right) and high (left) power photomicrographs with quick stain showing mixed stromal epithelial fragments with epithelial cells showing uniform nuclear size and chromatin. No feature of malignant changes was seen.

In conclusion, LGPA is an uncommon differential of a superotemporal mass, especially in children. Nevertheless, LGPA should be kept in mind when dealing with superotemporal masses in children. CT scan and FNAC help in ruling out other possible causes, each having diverse treatment approaches, for example, lymphoma validates an early institution of chemotherapy. This highlights the importance of CT scan and FNAC in achieving the accurate preoperative diagnosis and inducing the appropriate treatment without delay.

Learning points

- ▶ A high index of suspicion is necessary for diagnosing lacrimal gland pleomorphic adenoma in children less than 10 years.
- ▶ A CT scan aids in localisation and differentiating the tumour from a malignant lesion.
- ▶ Fine needle aspiration cytology aids in appropriate histopathological characterisation of the tumour and subsequent management.

Competing interests None.

Patient consent Obtained.

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