Cerebrospinal Fluid Rhinorrhoea Secondary to Amyloidosis of the Sphenoid Sinus

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SUMMARY

Amyloidosis of the skull base is a rare entity. A patient with localized amyloidosis of the sphenoid sinus presented at our institution with cerebrospinal fluid rhinorrhoea. Endoscopic excision of the lesion and multilayered obliteration of the sphenoid sinus resolved the symptoms.

KEY WORDS:
Sphenoid Sinus, Amyloidosis, Cerebrospinal Fluid Rhinorrhoea

INTRODUCTION

Amyloidosis is a broad term that describes diseases characterized by extracellular deposits of fibrillar proteins in tissues. In the head and neck region, larynx is the most commonly involved site[1]. Other sites include the tongue, oral cavity, salivary glands, nasopharynx and sinonasal regions. The sinonasal form usually presents as nasal obstruction, recurrent epistaxis, nasal blockage and conductive hearing loss secondary to Eustachian tube dysfunction[2,3].

CASE REPORT

A 48 years old, gentleman, was referred to our center, with cerebrospinal fluid leak from the left nostril for further assessment and management.

A CT-scan of paranasal sinuses wasdone, which showed a soft tissue mass in the region of the sphenoid body and sphenoid sinus extending posteriorly into the clivus associated with destruction of bones in this region. There was bony defect in the floor of the pituitary fossa, sphenoid sinus and roof of the nasopharynx. (Fig. 1)

He was started on intravenous ceftriaxone. Endoscopic examination under general anaesthesia was performed. Intraoperatively CSF leak was noted and a mass was seen in the left sphenoid sinus. Debulking of the tumor and a tissue biopsy was taken. The defect was repaired with abdominal fat, surgicel and gelfoam.

Histology of the biopsy tissue showed a few fragments of predominantly amorphous hyaline material, which were infiltrated by small numbers of chronic inflammatory cells (Fig. 2). In focal areas, the fragments show surface epithelial lining, composed of benign ciliated columnar epithelium. Congo-red positivity was demonstrated, with presence of green bifringence on cross polarized microscopy examination. A diagnosis of Amyloidosis was established. There was no evidence of recurrent CSF leak up to 12 months on follow-up.

DISCUSSION

Amyloidosis can be divided into systemic and localized disease. There are two major forms of systemic amyloidosis,

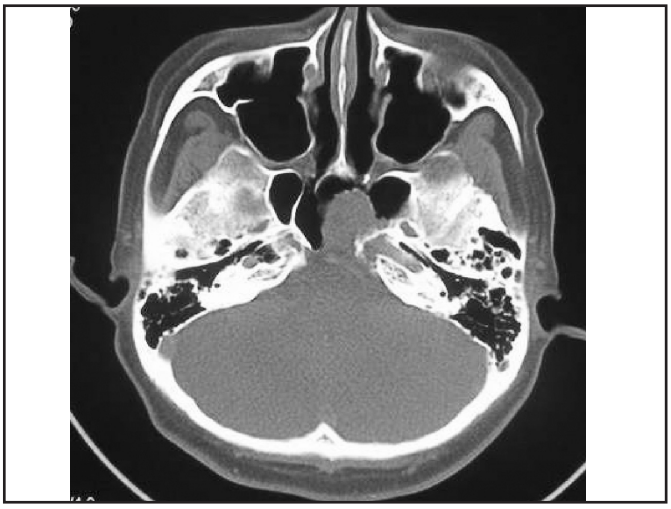

Fig. 1: Homogenous mass occupying the sphenoid sinus

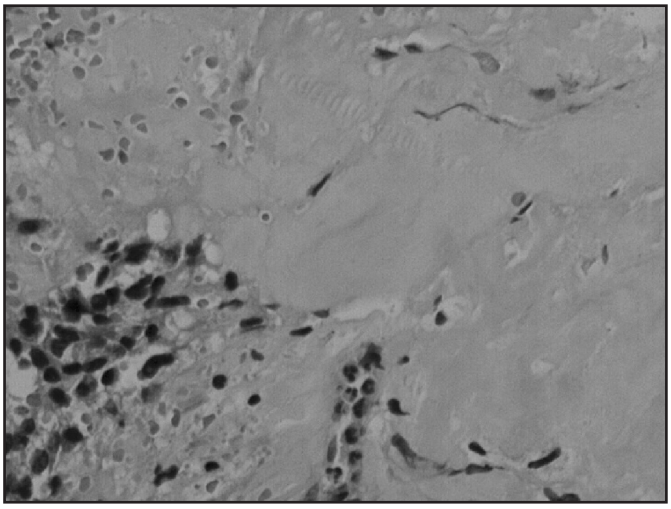

Fig. 2: Amorphous hyaline material with inflammatory cells

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AL and AA. AL amyloidosis is associated with primary systemic amyloidosis, multiple myeloma or plasma cell dyscrasias. AA amyloidosis or secondary amyloidosis is associated with chronic inflammatory conditions such as rheumatoid arthritis, Crohn's disease, inflammatory bowel disease and connective tissue disorders. Familial forms of amyloidosis have been described[4].

The new classification systems used today are based on the type of protein, which makes up the amyloid fibril. In the case of primary systemic, the fibrils are made up of light chain immunoglobulin produced by monoclonal plasmacytomas. In familial amyloidosis, these are strands of prealbumin. In secondary amyloidosis, a serum protein called serum amyloid A is produced. These excess strands then form relatively insoluble beta-pleated sheets, which are deposited in various tissues. These deposits then cause disease in various organs by interfering with their function.

In the head and neck region the localized form, AL type is most common. It is rarely associated with systemic involvement except for the tongue where amyloid induced macroglossia has a high association with plasma cell dyscrasias[5].

Amyloidosis of the head and neck is a benign disease with slow progression. The general consensus on management is conservative unless symptomatic. Local excision is the treatment of choice when necessary. Non-surgical treatment including chemotherapy, radiotherapy, local and systemic steroids has not been shown to be successful[3]. The primary aim of management in this case was to seal the cerebrospinal fluid leak and to prevent meningitis. The involvement of the skull-base as in this case, requires close follow-up and repeat surgery if necessary.

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