Case Report

A rare case of amyloidoma of parotid gland

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INTRODUCTION

Amyloidoma constitutes a solitary, localized, tumor-like deposit of amyloid, i.e. insoluble fibrillar proteins with beta-pleated sheet arrangement, in diverse organs without evidence of systemic amyloidosis.1 Amyloidosis is a heterogeneous group of disorders characterized by extracellular deposits of amyloid in individual organs or tissue. Extracellular amyloid consists of unique protein fibrils.2 Amyloidosis can be hereditary or acquired, and it may either be systemic or localized. We here report a rare case of amyloidoma confined to the parotid.

CASE REPORT

A 62 years old male patient with no comorbidities presented to ENT OPD with chief complaints of a painless swelling in the right parotid region of 7 months duration. No history of any previous trauma. On examination, there was a single 3x2 cm firm non tender well circumscribed swelling present in the right parotid region. There was no evidence of facial nerve paralysis or weakness and cervical lymphadenopathy.

Fine needle aspiration cytology findings were suggestive of spindle cell tumour.

Contrast-enhanced computed tomography (CECT) of neck reveals an approximately 25x25 mm sized minimally enhancing hypodense lesion with calcification in right superficial lobe of the right parotid gland suggestive of pleomorphic adenoma.

Surgery

Right superficial parotidectomy was done under general anesthesia on April 2019. Intraoperatively, facial nerve branches were seen going into tumor tissue, carefully dissected out and was sent for histopathological examination. Intraoperative and postoperative events were uneventful.

Histopathological examination

Surgical specimen sent was containing multiple grey brown soft tissue bits and the largest bit on cut section showed a homogenous grey white firm tumor measuring
2.8×1.5×1 cm along with surrounding normal salivary tissue. The growth was 0.2 to 0.6 cm from resected margin. On microscopic examination, there were normal salivary acini and ducts and the adjoining soft tissue shows deposition of acellular eosinophilic material with laminated bodies formation and foreign body giant cell reaction. On congo red stain, these eosinophilic material was congophilic and give apple green birefringent when seen under polarized light consistent with amyloid. On immunohistochemistry, SAA and lambda were negative and kappa was positive suggestive of amyloid tumor with kappa restriction.

Figure 1: (A) CECT neck axial view of the patient showing enhanced hypodence lesion in right superficial lobe of parotid, (B) CECT neck coronal view showing enhanced hypodense lesion in right superficial lobe of parotid.

Figure 2: Post-operative picture of patient.

Figure 3: Histopathological picture of patient suggesting amyloidoma.
Patient was investigated for systemic amyloidosis and multiple myeloma. Results of laboratory studies for systemic amyloidosis including a rectal biopsy were negative. A bone marrow biopsy, bone survey and serum and urine electrophoresis were negative for multiple myeloma and monoclonal gammapathy. A 6 months later the patient was fine without any clinical or laboratory evidence of systemic amyloidosis or multiple myeloma.

DISCUSSION

Amyloidosis was a heterogeneous group of disorders characterized by extracellular deposition of amyloid in individual organs or tissue. Extracellular amyloid consists of unique protein fibrils. Amyloidosis can be hereditary or acquired, and it may either be systemic or localized. The most frequent types of amyloidosis are the AL (primary) and AA (secondary) type. AA amyloidosis can develop in patients with chronic inflammatory conditions such as rheumatoid arthritis or chronic infections (e.g. osteomyelitis or tuberculosis). AL amyloidosis was due to deposition of protein derived from immunoglobulin light chain fragments, e.g. in multiple myeloma or Waldenström’s macroglobulinemia, with a monoclonal pattern on serum protein electrophoresis. Amyloid fibrils are composed of polypeptide fragments of normal serum proteins composed of kappa and lambda chains.

Amyloidoma or localized tumoral amyloidosis was the least common presentation of tissue amyloid deposition. It has been reported in many different anatomic regions, mainly the respiratory, genitourinary and gastrointestinal tracts, as well as the nervous system, breast, skin, cornea, vaginal mucosa and soft tissues. Amyloidoma of the parotid was a very rare condition, and only a few cases have been described in the literature so far. The diagnosis can be established with histologic examination of the involved tissue and, in particular, positive Congo red staining for amyloid.

The cause of the pathologic protein deposits was a monoclonal gammapathy proved by the evidence of ALκ amyloid, as confirmed in our patient by immunohistochemistry. The amyloid consists of monoclonal immunoglobulin-κ light chain that proves resistant to protein degradation and forms extracellular fibrillar, insoluble deposits with the known specific characteristic of amyloid (i.e. apple-green birefringence under polarized light). The monoclonal gammapathy may occur in a strictly localized manner and can be considered as an isolated form of plasmocytoma without spreading to other organs. The treatment of amyloidoma consists of complete surgical resection. In case of partial excision, recurrence was common. Long-term follow-up was required to rule out development of systemic disease. When localized in a parotid gland, as in the present case, the presence of amyloid may be a secondary manifestation of Sjogren’s (sicca) syndrome or possibly evidence of a primary lymphoma. The possibility of systemic amyloidosis and as a concomitant finding of multiple myeloma must be considered. The patient requires a thorough clinical and laboratory evaluation to support the diagnosis of AL amyloid tumor in any location.

CONCLUSION

This rare case of amyloidoma of parotid gland was successful treated by right superficial parotidectomy with facial nerve preservation. Till date only 1 case have been reported so far of amyloidoma in head and neck region. This case was being reported due to rarity of its presentation.

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