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Giant primary amyloidoma of the tonsil

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Abstract

A case of giant amyloidoma in the left tonsil with extensive osseous metaplasia and a scanty and patchy monoclonal population of IgG Kappa plasma cells, is presented.

Localized tumoral amyloidosis is a rare, benign tumour of the upper aerodigestive tract. Organ-limited amyloidosis has been shown to be confined to various systems but, since the lesion was first described, only four cases situated in the tonsils have been reported in the English literature, and none of these had either osseous metaplasia or a monoclonal population of IgG Kappa plasma cells.

Key words: Tonsillar neoplasms; Amyloidosis

Introduction

The term 'amyloidosis' includes a diverse collection of diseases characterized by the presence of amorphous extracellular eosinophilic deposits of unique protein fibrils that give apple-green birefringence under polarized light after staining with Congo red (Krishnan *et al.*, 1993).

Amyloidomas or localized tumour-like amyloid deposits have been observed in a variety of locations in the head and neck, mainly in the larynx (McAlpine and Fuller, 1964) and thyroid (Michaels and Hyams, 1979), but are extremely rare in the tonsils.

At present only four previous reports have been published in the English literature of amyloid deposits located in the tonsils (Eriksen, 1970; Benjamin, 1977; Michaels and Hyams, 1979; Beiser *et al.*, 1980). In addition, we found four other reports in the non-English language (Mutschler, 1933; Chiari, 1937; Marco, 1985; Lauritzen, 1987) (Table I).

In this paper we present one case of amyloidoma located in the left tonsil. We review the literature on this tumour and discuss the clinical features, histopathology and evolution.

Case report

A 72-year-old Caucasian male was admitted to the Otolaryngology department of 'Juan Canalejo' Hospital (La Coruña; Spain) for evaluation of a left tonsil enlargement first noticed five years earlier. The patient initially consulted his family physician but no further investigations were made.

His past medical history was unremarkable apart from high blood pressure. There were no previous tonsillar infections or other symptoms. Family history was also negative.

On admission the physical examination revealed a 5 × 4 cm painless left tonsil. Its consistency was firm and uniform, the surface was smooth and the margins were definite. There was neither clinical evidence of deep

TABLE I

REPORTED CASES

Authors	Year	Age	Sex	Sites of involvement	Symptoms	Comments
Multscher	1933	35	F	Lower pole right tonsil	Asymptomatic	First case (Italian report)
Chiari	1937	58	F	Almost complete left tonsil	Asymptomatic	German, non-English, report
Eriksen	1970	59	M	Both tonsils	Dysphagia and dyspnoea	Severe obstruction
Benjamin	1977	54	M	Nodule of left tonsil	Asymptomatic	
Michaels and Hyams	1979	28	M	One palatine tonsil	Discomfort in the throat	Report of 25 respiratory cases
Beiser *et al.*	1980	35	M	All Waldeyer's ring	Discomfort in the throat	Involves all ring's structures
Marco	1985	67	F	Both tonsils	Dysphagia and sore throat	Spanish, non-English, report
Lauritzen	1987	50	M	One complete tonsil	Dysphagia and sore throat	Danish, non-English, report
Present study	1996	72	M	Complete left tonsil	Discomfort in the throat	Old man

NON-ACCEPTABLE CASES

Authors	Year	Age	Sex	Sites of involvement	Comments
McAlpine *et al.*	1963	54	F	Polypoid structure in left tonsil	Amyloid was not confirmed on further histological investigation.

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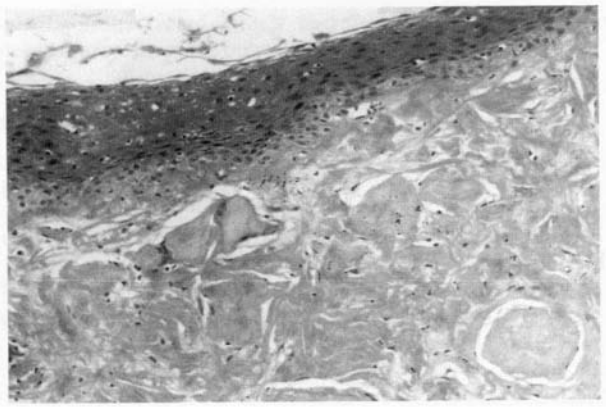

FIG. 1

Large deposits of amyloid are present under the surface of the tonsillar epithlium.

infiltration nor lymphadenopathy. The remainder of his head and neck examination, including the contralateral tonsil, nose, nasopharynx, tongue base, larynx and neck were normal.

Chest X-ray, urine analysis, including microscopic examination of sediment, and complete blood count, with differential, electrolytes and blood levels of glucose, urea, uric acid, albumin, globulins, creatinine, bilirubin, alkaline phosphatase and SGO transaminase, were all within the normal range. Erythrocyte sedimentation rate was elevated at 48 mm/hour (Normal range 0–20).

A left tonsillectomy was performed under general anaesthesia. The tonsil was entirely occupied by an ovoid mass that measured 4.5 × 4 cm and had a stoney consistency and bony spiculae visualized macroscopically on the cut surface.

The microscopic study revealed large, homogeneous, amorphous, eosinophilic deposits obliterating the tonsillar structures and infiltrating the neighbouring tissues (Figure 1). This material showed a positive staining with the Congo Red stain and an apple green birefringence under the polarizing lenses. When the slides were pretreated with potassium permanganate the Congo Red positivity remained essentially unchanged.

A foreign body giant cell reaction around the amyloid masses (Figure 2) and focal areas of bony metaplasia, some of them with bone marrow (Figure 3), admixed with the amyloid were present. The lymphoid tissue of the tonsil was inapparent and isolated focal collections of typical plasma cells were present around blood vessels. Immunostaining in paraffin embedded sections for heavy and light immunoglobulin chains showed a IgG-Kappa monoclonal component.

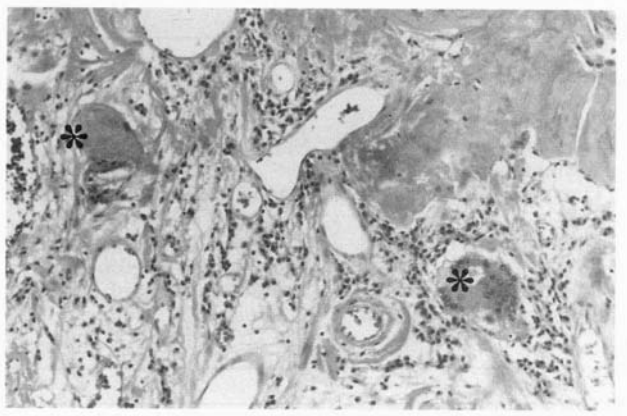

FIG. 2

Foreign body giant cell reaction (asterisk) and cytologically normal plasma cells are present around blood vessels within the amyloid.

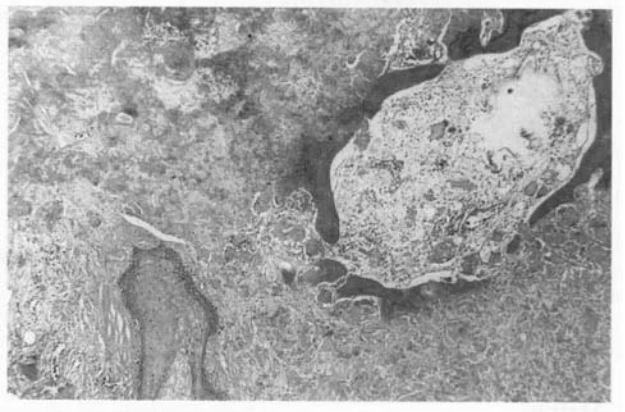

FIG. 3

Bone formation within the amyloid and adjacent to the epithelium of a crypt.

Post-operative healing was uneventful. Further evaluation by the Division of Haematology and Internal Medicine did not reveal any systemic disease. Measurement of blood immunoglobulins showed normal levels and pattern. Serologic tests: VDRL, rheumatoid factor, antinuclear factor, monotest, and serum calcitonin were negative. Rectal, tongue and lip biopsies were all negative for amyloid deposits; and bone marrow biopsy was normal. Urine protein immunoelectrophoresis was negative for Kappa Bence-Jones proteins. Imaging studies, including echocardiogram, radiological skeletal survey and thorax CT scan, and electromyography and motor and sensorial conductive velocity were also within the normal range.

The patient has been followed at regular intervals for four years and there has been no evidence of systemic amyloidosis nor multiple myeloma development.

Discussion

Since Rokitansky, the first to describe systemic amyloidosis in 1842, and Virchow who coined the word amyloid in 1853 (Mitrani and Biller, 1985) numerous classifications of amyloidosis have been proposed. Subdivision today is based on the chemical type of proteins that forms the fibrils, type 'AA' (acute phase reactant serum amyloid A protein), type 'AL' (light amyloid protein) chains of immunoglobulins amyloid protein (Glenner, 1980a, b).

There are three types of systemic amyloidosis: (1) immunocytic dyscrasia associated, characterized by 'AL' proteins; (2) reactive, which contains 'AA' protein, and (3) heredofamilial forms, composed of different proteins, such as transthyretin, gelsolin, cystatin-C or AA (Krishnan *et al*, 1993).

The term localized refers to an amyloidosis that affects a single organ, without evidence of generalized involvement and could be classified as AL and/or AA types (Jensen *et al*., 1985). Most of the cases of primary localized amyloidosis have been reported in the upper and lower respiratory tracts (Benjamin, 1977).

Perusal of the available literature disclosed two types of localized amyloid deposit in tonsil, patients of Chiari (1937), Eriksen, (1970), Lauritzen (1987) and our report had a real amyloidoma (amyloidosis manifested as a localized mass). However, Marco's (1985), Beiser's (1980), Multschler's (1933) and Benjamin's (1977) cases present amyloid nodules or plaques on the surface of the tonsils. The case reported by Michaels and Hyams (1979) had few details of the lesion.

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Review of the previous reported tonsillar cases and the present study reveals an age range from 28 to 72 years, with a mean of 50.89 years. However, our patient was the oldest of the series in which 66.67% of patients were male.

We have not included the case reported by McAlpine *et al.* (1963) because there are serious doubts about the diagnosis and amyloid was not confirmed on further histological investigation.

Localized amyloidosis symptoms, when present, are caused by the physical presence, size and location of the deposits (Simpson *et al.*, 1984). In most of the cases patients were asymptomatic or had small complaints, as in our case. Three cases of tonsillar amyloidosis have been reported to present with a globus sensation and dysphagia and one patient had a severe airway obstruction, secondary to a very large tonsil enlargement (see Table II).

The diagnosis is by tissue biopsy, the treatment is surgical removal and the prognosis is very good (Simpson *et al.*, 1984).

In all patients with localized amyloidosis it is mandatory to rule out the presence of systemic involvement (Clevens *et al.*, 1994). The importance of determining whether amyloid deposits represent localized or systemic amyloidosis was pointed out by Kyle and Bayrd (1975) who stressed the markedly shortened life expectancy in systemic forms of amyloidosis. In our patient all further investigations were negative.

Our case presents two unusual histological features: it is unique in that osseous metaplasia, with trabeculae of bone, is reported. This appearance is similar to the one of the tracheopathia osteoplastica, a rare entity that is associated with the presence of osseous metaplasia and, frequently, amyloid deposits in the trachea (Ashley, 1970). On the other hand, a rather scanty monoclonal population of IgG Kappa plasma cells was demonstrated. Differential diagnosis with plasmocytomas of the respiratory tract depends on the number and type of plasma cell. In plasmocytoma the mass is composed almost entirely of neoplastic plasma cells (Hyams *et al.*, 1988), and sometimes is complicated by the formation of amyloid deposits (Michaels and Hyams, 1979) In amyloidomas, however, the infiltrate of plasma cell is small and no atypia is present. Nevertheless, the monoclonality of this sparse population (IgG-Kappa) makes mandatory the close follow-up to rule out the development of a multiple myeloma.

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