CASE STUDY

Basilingual Xanthoma

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Abstract Xanthomas are benign slow-growing dysmetabolism tumours consisting of histiocytes laden with lipid droplets of cholesteryl ester and, sometimes, triglycerides. Two forms of appearance have been identified: as dyslipidemic xanthomatosis with coexisting alteration of lipids, or as normolipidemic xanthomatosis without any lipid level alterations.

Involvement of the upper area of aerodigestive tract by this entity is very rare and there are only a few cases in the literature.

We report a case of dyslipidemic xanthomatosis with epiglottic and basilingual involvement that produced oropharyngeal foreign body sensation with dysphagia. Evolution was favourable after laser resection.

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PALABRAS CLAVES
Xantoma basilingual

Resumen Los xantomas son tumores benignos dismetabólicos de crecimiento lento constituidos por histiocitos cargados de pequeñas gotas lipídicas de colesterol esterificado y, a veces, triglicéridos. Se han individualizado dos formas de aparición: como xantomatosis dislipémica en la que coexiste una alteración de los lipidos, o bien como xantomatosis normolipémica.

La afectación de las vías aerodigestivas superiores por esta entidad es rara y hay pocos casos recogidos en la literatura.

Presentamos un caso de xantomatosis dislipémica con afectación basilingual y epiglotítica que debutó con sensación de cuerpo extraño orofaringeo y disfagia y evolucionó favorablemente tras la resección con láser.

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Case Report

We present the case of a 40-year-old male with a family history of dyslipidemia and ischaemic heart disease at an early age. The personal history included dyslipidemia and tonsillectomy, with no toxic habits.


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The patient attended consultation due to dysphagia for solids and liquids, foreign body sensation and chronic and progressive snoring of 3 months evolution. These symptoms were accompanied by guttural voice and breathing difficulty which disappeared in the lateral position, as well as subjective and unquantified sensation of weight loss.

Examination by anterior rhinoscopy revealed a left septal deviation. No alterations were found in the oropharynx or neck. Indirect laryngoscopy also revealed a large mass located at the level of the left tongue base which extended to the vallecula, clearly exceeded the midline and pushed the epiglottis, preventing visualisation of the vocal cords. Fibroscopic examination revealed no additional abnormalities in the larynx and hypopharynx.

We decided to hospitalise the patient for treatment and study. He experienced clinical improvement and partial reduction of tumour size with glucocorticoid treatment during the 10 days of admission. We took 4 samples of the tongue base, measuring between 0.3 and 1 cm in diameter, whose anatomopathological result was of squamous mucosa with moderate, focally acute, chronic inflammation and presence of PAS+ microorganisms, histologically compatible with fungi. No metaplastic cells were located.

The patient underwent an endocrinological study which discovered hypercholesterolemic and hypertriglyceridemic mixed dyslipidemia (total cholesterol [TC]: 280 mg/dL; low-density lipoprotein [LDL]: 79 mg/dL; very-low-density lipoprotein [VLDL]: 44 mg/dL, and triglycerides [TGC]: 220), whose control through dietary measures resulted in a normalisation of cholesterol levels, albeit with an increase in triglyceride levels (TC: 231 mg/dL; LDL: 107.7 mg/dL, and TGC: 316). We did not observe any alterations in the liver, thyroid or immune system.

A magnetic resonance imaging (MRI) study (Fig. 1) confirmed the presence of a 38 mm × 35 mm × 32 mm mass at the level of the tongue base, predominantly on the left side, which infiltrated the tongue base and decreased the calibre of the airway. The lesion was hypointense on T1 sequences, discreetly hyperintense on T2 sequences and showed contrast uptake. In addition, there was a right lateral adenopathy of 1 cm and several small adenopathies in the left laterocervical chain, all of unspecific character.

The patient reported clinical improvement, but given the persistence of the tumour mass during the exploration and the anatomopathological findings, we decided to perform surgical excision of the lesion. After general anaesthesia

![MRI image](image_url)
and orotracheal intubation we observed a thickening of the tongue base and a yellowish mass which extended from the lingual surface of the epiglottis towards the tongue base by the left vallecula, clearly exceeding the midline. This was resected with a 4-W diode laser in superpulse mode, respecting the epiglottic cartilage.

The pathological study (Fig. 2) through haematoxylin–eosin staining showed the presence of foamy histiocytes, Touton cells and chronic inflammation, consistent with xanthoma. The immunohistochemical study revealed positivity for CD 68 marker.

The postoperative course was favourable and without complications. Imaging follow-up through computed tomography (CT) at 12 months, and clinical follow-up through periodic fibroscopies for 18 months have not identified lesion recurrence. The patient has remained asymptomatic.

Discussion

Xanthomatosis is an uncommon entity which usually affects the skin and is characterised by the appearance of aggregates of foamy macrophages or lipid-laden histiocytes which are macroscopically manifested as yellowish, brownish or orange multifoci of plaques, nodules or papules, exhibiting a slow and benign growth. These lesions are known as xanthomas and have different morphologies depending on the alteration causing them: eruptive xanthomas in hypertriglyceridaemia and diabetes with poor metabolic control; xanthelasmas in hypoproteinemia; tuberous in hypercholesterolemia; plane in myeloma.¹

There are 2 forms of appearance: as normolipemic xanthomatosis or else as dyslipidemic xanthomatosis. In cases of dyslipidemic xanthomatosis there is an alteration of circulating lipids, either idiopathic or secondary to diabetes mellitus, pancreatic disorders, hypothyroidism, biliary obstruction or genetic syndromes such as von Gierke disease.¹² In the case of normolipemic xanthomatosis, the formation of xanthomas is secondary to an increase in the formation of intracellular lipids of unknown aetiology.

The anatomopathological study shows infiltration of lipid-laden foamy cells and giant cells called Touton cells, characterised by having multiple nuclei arranged in an incomplete circle.¹

The prognosis of xanthomatosis is favourable. Xanthomas generally disappear after the normalisation of blood lipid levels by dietary restriction.¹³ The main prognostic factor is the presence of vascular atherosclerosis and the level of cardiac involvement.¹³

Xanthomas usually have a slow growth and are non-invasive, although it is true that they may produce local functional involvement which, depending on the affected area, may require surgical resection. Furthermore, xanthomas often recur when those are not completely removed. Radiotherapy has demonstrated efficacy in treating xanthomatosis.¹³⁴

There are few reports in the literature affecting the ENT area.¹

A case of xanthoma at the level of the posterior pharyngeal wall and extending from the hypopharynx to the nasopharynx was described in a patient with hypercholesterolemia and disseminated xanomas.³ The patient had a good response to therapy with atromid-S and surgical excision. Another case was described located in the left posterior pharyngeal wall in a normolipemic patient.⁴ The evolution after surgical resection was favourable.

Involvement of the paranasal sinuses has also been reported in very few cases: Raymond⁵ and Malik et al.⁶ described cases of maxillary involvement in normolipemic patients who presented prior cutaneous xanthomas.

Multifocal lesions located at the level of the tongue, epiglottis, right vocal cord, tonsils and mouth have been described by Travis et al.⁷ and in another case by Fernández Pérez et al.⁸ The latter patient was normolipemic and presented xanthomatous plaques in the vallecula, right ventricular band and anterior laryngeal commissure.

Szyfter et al.⁹ described a case of xanthomatosis with cutaneous involvement, subglottic stenosis and mesopharyngeal plaques which were stabilised through pharmacological treatment with prednisone and amiodipine.

The current case was a dyslipidemic patient who presented a single lesion located in the vallecula, and affecting the lingual surface of the epiglottis and the lingual tonsil.
In conclusion, this is a rare nosological entity in the area of the upper aerodigestive tract. It is benign and has a good response to treatment. This entity should not be left out from the differential diagnosis of patients presenting a mass in the ENT area.

Conflict of Interest

The authors have no conflict of interest to declare.

References


