

Cutaneous Pleomorphic Adenoma or Chondroid Syringoma: A Case Report

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Abstract

Case Report

Cutaneous pleomorphic adenoma or mixed skin tumor or chondroid syringoma is an exceptional benign tumor of the head and neck region. We present in this article a new case of chondroid syringoma of the left nasolabial fold evolving for 5 years in a 69-year-old patient, with no particular pathological history. The tumor was completely resected under local anesthesia and its anatomic-pathological examination had confirmed the diagnosis of mixed skin tumor. Long-term follow-up did not show a recurrence after 2-years. Through this clinical observation and a review of the literature, we will discuss epidemiological, clinical, histological, therapeutic features of this exceptional tumor.

Keywords: chondroïde syringoma, face, histology, surgery.

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INTRODUCTION

Chondroid syringoma or cutaneous pleomorphic adenoma is a rare benign skin tumor that forms a subcutaneous or intradermal nodule in the head and neck area [1]. Its diagnosis is sometimes difficult and often overlooked by pathologists; these tumors are characterized by a double epithelial and mesenchymal component [2]. We report a new observation of chondroid syringoma of the face occurring in a 65 year

old man and we discuss through a literature review the different features of this rare skin tumor.

OBSERVATION

This is a 69-year-old mountain man with no particular pathological history who presented to the otorhinolaryngology consultation of the military field hospital for a swelling of the left nasolabial fold evolving for 05 years without associated clinical signs apart from a psychic and aesthetic impact (fig. 1).



Fig-1: Exuberant tumor of the left nasolabial fold

The tumor started with a small, painless papule which progressively increased. The ENT clinical examination revealed a superficial, hard, painless and irreducible swelling of the left nasolabial fold, with a form of a smooth dome, not exceeding 2cm at the major axis. The otological, rhinological and oropharyngeal examination did not find any other mass or anomaly. The palpation of the lymph nodes was without particularities. A paraclinical checkup was not requested because of the lack of resources in a field hospital.

The patient benefited, under local anesthesia, from a surgical intervention which allowed the complete excision of the tumor with safety margins of 0.5cm. Dissection of the tumor was easy and the closure was made by bringing the edges of the incision into two planes. (fig. 2-3).



Fig-2: Complete removal of the tumor



Fig-3: Skin closure in two planes

The anatomico-pathological examination of the operating specimen confirmed the diagnosis of mixed skin tumor by showing a tumor proliferation limited at the periphery by fibrous condensation. It was made up of an epithelial and mesenchymal component. The epithelial one was made up of irregular spans, tubes, pads and acini made up of small cells with regular nuclei. There were squamous foci metaplasias. These

epithelial elements, sometimes grouped in range, were immersed in a hyaline myxoid substance. The excision was complete, but focally economic (fig. 4).

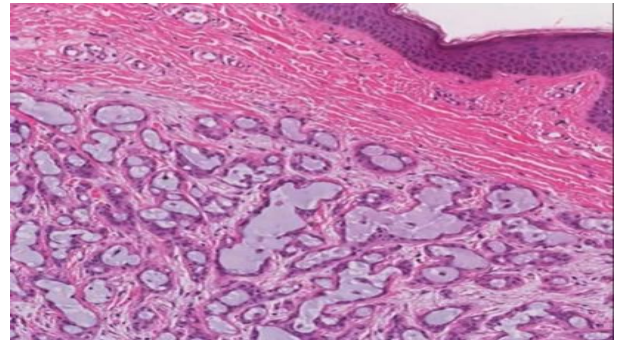


Fig-4: histological section of the tumor showing tumor proliferation limited at the periphery by fibrous condensation

The immediate post-operative follow-up was simple, and long-term monitoring did not show recurrences after 2 years of follow-up.

DISCUSSION

Billroth in 1859 first described chondroid syringoma and initially named it "mixed skin tumor" by analogy to mixed tumors of the salivary glands [3]. In 1961, Hirsch and Helwig called it "chondroid syringoma" because of the presence of a "cartilage-like" stromal component and sweat glands [3].

This tumor affects often males at a median age of 50 years with extremes ranging from 22 to 73 years [4]. The sex ratio varies between 1.3/1 and 5/1. It occurs mainly in the head and neck (85% of cases) with a predilection for the nose, upper lip and eyebrows [5]. Other areas of the body may be affected: the shoulder, leg, fingers or toes [5].

Clinically it is a slow growth, firm and painless dermic and / or hypodermic nodule, measuring between 0.5 and 3 cm in diameter, usually not adherent to the deep subcutaneous planes [6]. The clinical differential diagnosis is mainly made with basal cell carcinoma, sebaceous cyst, dermoid cyst, schwannoma, neurofibroma or pilomatricoma [3].

Histologically, the presence of the epithelial and mesenchymal contingents, characteristic of mixed tumors, allows the diagnosis of chondroid syringoma. The mesenchymal contingent can be myxoid, chondroid, adipose or fibrous and contains clusters of epithelial cells without glandular structure or isolated cells. The epithelial contingent is made up of glands whose histological appearance allows two types to be identified: the apocrine type, as in our case, is the most common and is made up of tubular or cystic glands, focally branched, bordered by a double layer of cuboid or flattened cells and the eccrine type characterized by narrower lumens and glands bordered by a single cell layer [7].

The immunohistochemical study shows that the cells bordering the lumens express the epithelial markers in favor of an adnexal origin: EMA, the cytokeratins CK7, CK14 and ACE [8,9]. The cells not bordering the lumens of the epithelial mass show a labeling with the antibodies of the myoepithelial cells, PS100 and p63. Other markings have been reported: GFAP (glial fibrillary acidic protein), NSE [2].

The origin of this tumor remains under discussion. The hypothesis talk about an origin from the secretory and ductal elements of the sweat glands, which would explain the positivity of p63 and p504s observed in same cases. More recently Kazakov *et al.*, in a series of 244 cases, have reported that this tumor is more of an apocrine nature [10]. The majority of chondroid syringomas is benign, but malignant forms, although very rare, may exist [11]. These malignant tumors latter are either primitive or degenerating after a very long evolution or incomplete resection [5].

The treatment of choice is based on wide excision with safety margins of 3 to 4 mm in healthy tissue. Dissection is often easy due to the existence of a cleavage plane and the proper delimitation of the lesion by a capsule. Complete excision is the only guarantee of the absence of recurrence [4]. In our case, the entire lesion was excised. No recurrences have been noted after two years of follow-up. Benign incompletely resected tumors require periodic follow-up to detect recurrence or possible malignant transformation [3]. The prognosis for this type of tumor is good.

CONCLUSION

Cutaneous pleomorphic adenoma or chondroid syringoma is a rare benign skin tumor. Unlike salivary localization, this tumor affects men more than women and often occurs in the cervico-facial region. It is difficult to evoke it clinically because of its rarity and the absence of specific signs, its diagnosis is purely histological. Its treatment is surgical and requires complete excision, the only guarantee to avoid recurrences and possible malignant degeneration. The prognosis for chondroid syringoma of the face is very good.

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