

CASE REPORT

Subcutaneous benign fibrous histiocytoma of the cheek. Case report and review of the literature

Fibroma istiocitario sottocutaneo benigno della guancia. Presentazione di un caso e revisione della letteratura

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SUMMARY

Fibrous histiocytoma is a benign tumour composed of a mixture of fibroblastic and histiocytic cells. Based on the location of this tumour, fibrous histiocytoma are usually divided into cutaneous types and those involving deep tissues. These lesions most often arise on the skin, but may rarely occur in soft deep tissues. The diagnosis of fibrous histiocytoma may be clinically difficult when the lesion is located in the deep tissues, and is frequently confirmed after local excision. The most important diagnostic distinction is the separation of this tumour from aggressive forms of fibrohistiocytic neoplasms, including dermatofibrosarcoma protuberans and malignant fibrous histiocytoma. A 19-year-old male presented with a painless swelling on the right cheek. Detailed clinical and laboratory examinations were performed. The lesion had been totally excised under local anaesthesia, and histopathology revealed a benign fibrous histiocytoma. The diagnosis, location, treatment and prognosis of fibrous histiocytoma are also discussed.

KEY WORDS: Benign tumour • Cheek • Benign Fibrous Histiocytoma

RIASSUNTO

Il fibroma istiocitario è un tumore benigno composto da fibroblasti e istiociti. In base alla sede di questo tumore, i fibromi istiocitari sono divisi solitamente nei tipi cutanei ed in quelli che coinvolgono i tessuti profondi. Queste lesioni si presentano spesso sulla cute, mentre possono riscontrarsi raramente nei tessuti molli profondi. La diagnosi clinica del fibroma istiocitario può essere difficile quando la lesione è situata in profondità e frequentemente se ne ha conferma solo dopo l'asportazione. La distinzione diagnostica più importante è la differenziazione di questo tumore dalle forme aggressive del tumore fibroistocitario, compreso il dermatofibrosarcoma protuberans e l'istiocitoma fibroso maligno. Caso clinico: un paziente maschio di 19 anni si presenta con tumefazione non dolorosa alla guancia destra. Vengono effettuati gli esami di laboratorio e clinici. La lesione viene completamente asportata in anestesia locale. L'esame istologico ha rivelato un istiocitoma fibroso benigno. Sono discussi la diagnosi, la sede, il trattamento e la prognosi del fibroma istiocitario.

PAROLE CHIAVE: Tumori benigni • Guancia • Istiocitoma fibroso benigno

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Introduction

Fibrous histiocytoma (FH) is a benign tumour composed of a mixture of fibroblastic and histiocytic cells. This tumour most frequently occurs in the dermis, but is also sporadically found in soft tissue and parenchymal organs. The benign FH usually originates in sun-exposed skin and in orbital tissues, whereas the occurrence of this lesion in deep soft tissues of the head and neck has rarely been reported. The term cutaneous FH is usually used to refer to all superficial tumours of skin regardless of appearance. Similar lesions involving subcutis or deep structures will only be referred as FH^{1,2}. These neoplasms should be differentiated from the malignant FH, which frequently has a rather aggressive malignant course^{3,4}.

In the present study, a case with subcutaneous FH on the cheek is described. From a thorough search of the literature,

only 6 cases with subcutaneous benign FH of the head and neck area were found, only two of which located on the cheek^{1,2,5} (Table I).

Case report

A 19-year-old male presented in our ENT outpatient clinic, with an apparently painless swelling on the right cheek, complaining of a lesion that appeared 4 years previously. His past medical history was negative, apart from mild acne compatible with his age.

The clinical examination revealed a mass about 3 x 4 cm on the right cheek, of skin colour and hard consistency, which was not attached to the subcutaneous tissue or to the skin. The lesion had been increasing slightly in size over the last 6 months. Small, painless, mobile lymph nodes, on the right submandibular triangle, were also palpable.

Table 1. Review of literature of subcutaneous fibrous histiocytomas of head and neck.

No.	Ref.	Site	Age yrs	Sex	Treatment	Follow-up, outcome
1.	Fletcher, 1990	Subcutaneous face	45	M	LE*X2	†Re at 2 Y ‡NED after 8Y
2.	Fletcher, 1990	Intramuscular scalp	31	M	LE	9 M, NED
3.	Fletcher, 1990	Subcutaneous scalp	58	F	LE	10 M, NED
4.	Fletcher, 1990	Subcutaneous scalp	10	F	LE	**NA
5.	Fletcher, 1990	Subcutaneous cheek	60	M	LE	NA
6.	Fletcher, 1990	Intramuscular cheek	56	M	LE	NA
7.	Our case	Subcutaneous cheek	22	M	LE	4 Y, NED

* LE: Local excision; † Re: recurrence; ‡ NED: no evidence of disease; ** NA: information not available

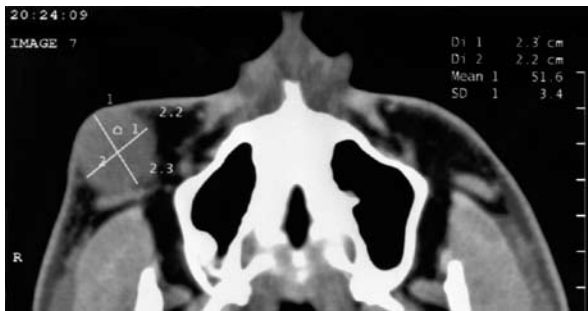


Fig. 1. CT scan shows homogenous enhancement of mass on right cheek.

Full blood count, erythrocyte sedimentation rate, renal urea and creatinine, as well as, liver tests were all within normal limits. Echography and Doppler echography of the mass suggested a lesion of vascular origin. A computed tomography scan was performed with administration of intravenous contrast, which revealed a mass of soft tissue, the dimensions of which were 3 x 3.5 cm (Fig. 1). The lesion had well-defined borders, not infiltrating the surrounding tissues. These radiologic findings were not compatible with the diagnosis of a lymph node, but were rather indicative of a benign vascular lesion.

The lesion, of whitish colour and of fibroelastic consistency, had been totally excised under local anaesthesia. Pathology examination revealed a neoplasm of mesenchymal origin, rich in cells with morphology and immunohistochemical characters compatible with benign fibrous histiocytoma (Fig. 2). Immunohistochemical staining demonstrated positivity for CD68 (monoclonal, clone PG-M1 dilution 1:50, Dako) (Fig. 3A) and Vimentin (monoclonal, clone V9, dilution 1:50, Dako) (Fig. 3B), and negativity for smooth muscle actin (SMA) (monoclonal, clone 1A4, dilution 1:50, Dako) (Fig. 3C), S-100 protein (polyclonal, dilution 1:400, Dako) (Fig. 3D), CD34 (monoclonal, clone QBend 10 dilution 1:50, Dako) and factor XIIIa (polyclonal, dilution 1:200, Biocave). Microscopic and immunohistochemical

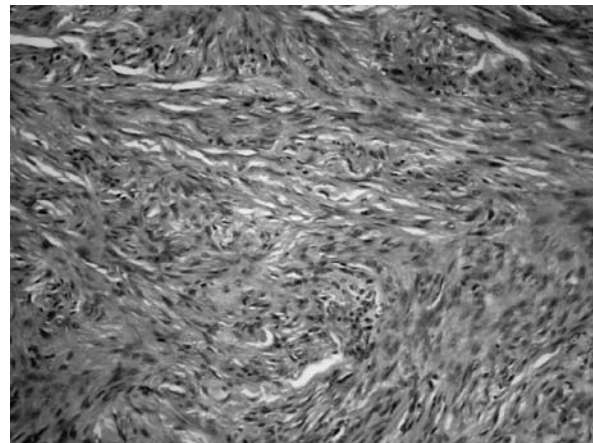


Fig. 2. Irregularly arranged collagen fibres mixed with scattered proliferating fibroblasts (Histiocytoma, fibrous type), X200, H + E.

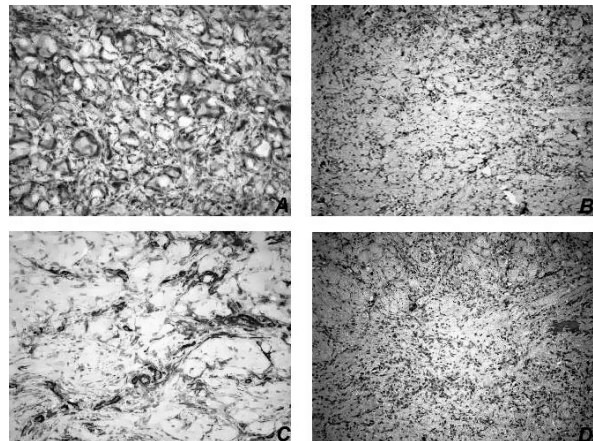


Fig. 3. Immunohistochemical staining demonstrated positivity for CD68 (A) and Vimentin (B), and negativity for smooth muscle actin (SMA) (C) and S-100 (D).

examinations confirmed the diagnosis of benign fibrous histiocytoma. The patient is asymptomatic with no evidence of recurrent disease after a 4-year follow-up.

Discussion

Benign FH was not known as a clinical entity before 1970 when, as a result of the development of immunohistochemical techniques and electronic microscopy, differential diagnosis became feasible^{6,7}.

The diagnosis of FH may be difficult clinically when the lesion is located in the deep tissues, and is frequently confirmed after local excision. Histopathologically, this tumour is a neoplasm of histiocytic origin and is composed of a biphasic cell population of histiocytes and fibroblasts⁸. This accounts for the dual population of histiocytic and fibrous elements commonly seen in this tumour. According to other Authors, the histiocyte originates from an undifferentiated mesenchymal stem cell⁹. In our case, immunohistochemistry was performed for differential diagnosis, showing similar features on microscopic examination. The positivity for CD68 and vimentin demonstrated that the lesion was composed of histiocytic cells and fibroblast-like cells on immunohistochemistry, and the negativity for SMA and S-100 could differentiate the lesion from leiomyosarcoma and neurogenic tumours¹⁰.

The most important diagnostic distinction is the separation of this tumour from aggressive forms of fibrohistiocytic neoplasms, including dermatofibrosarcoma protuberans and malignant FH^{11,12}. As with benign FH, the diagnosis of malignant FH frequently relies upon immunohistochemistry and electron microscopy to differentiate it from other lesions.

Malignant FH is composed of malignant pleomorphic sarcomatous cells, bizarre giant cells, and frequent mitotic figures¹³. Histologic features, unfortunately, play a minor role in predicting the biologic behaviour of these neoplasms. The presence of certain atypical histological features, including necrosis, marked cellular and mitotic activity, does not correlate well with clinical recurrence^{3,11,14}.

The difference between benign and malignant FH is usually obvious, because the latter is a pleomorphic, deeply situated tumour with numerous typical and atypical mitotic figures and prominent areas of haemorrhage and necrosis. Less obvious, however, is the difference between this tumour and the angiomatoid form of FH. The latter is a tumour occurring usually in childhood characterized by sheets of histiocytic cells interrupted by cystic areas of haemorrhage. These are surrounded by a dense cuff of lymphocytes and plasma cells, but hardly ever possess giant cells and xanthoma cells like the FH^{11,14,15}.

Like FH, dermatofibrosarcoma protuberans occurs in the dermis and subcutis, but is more apt to show extensive subcutaneous involvement than benign FH. It is also characterized by a more uniform cellular population and lacks giant cells, inflammatory cells, and xanthomatous elements. Its fascicles, composed of slender attenuated cells, are longer and arranged in a distinct storiform pattern, unlike the short curlicue fascicles of FH. Its margins are infiltrative in contrast to the well-defined margins of FH. Immunostaining also reveals distinct differences in the cellular composition

of these tumours. Fibrous histiocytomas contain a significant population of factor XIIIa-positive cells, although it has been debated whether these cells represent a population of neoplastic cells or a peculiar infiltrate that accompanies the tumour. In contrast, dermatofibrosarcoma protuberans contains only scattered factor XIIIa-positive cells, but, in striking contrast to benign FH, it expresses CD34 in a significant portion of neoplastic cells. The combination of these two stains has proved to be highly reliable in distinguishing these two lesions, which often cause diagnostic problems, particularly when only a superficial biopsy specimen is available for assessment^{12,14-16}.

FH, based on the location, are usually divided into cutaneous types and those involving deep tissues^{2,5}. These lesions usually arise on the skin, but may very occasionally occur in soft deep tissues¹⁶. Non-cutaneous benign FH represents approximately 1% of all benign FH lesions. This tumour has been associated with a previous trauma, sun exposure, and chronic infection, rather suggesting that it represents a reactive proliferation of benign cells. Benign FH of the non-cutaneous soft tissues of the head and neck most often develops as a painless mass with specific symptoms caused by interference with the normal anatomy and physiology of the area in which they arise¹¹.

Specific sites of involvement, on the head and neck region, described in the literature, include buccal mucosa, submandibular triangle, oral tongue, larynx, nasal cavity, mandible and supraclavicular fossa^{1,5,6,11,17,18}. Usually, these lesions appear very well circumscribed (often being encapsulated) and the cut surface is firm and pale to yellowish-brown in colour. There is no macroscopic evidence of necrosis but sometimes the lesions are focally cystic, presenting haemorrhage within. The maximum diameter has been reported to range from 2 to 12 cm⁵. It is worthwhile pointing out that the majority of lesions had only been shelled out, a fact that almost certainly contributed to the high local recurrence rate¹. All lesions proved to be solitary, none being related to a joint. Clinically, most lesions presented as a painless, slowly growing mass, the pre-operative duration ranging from 3 to 12 months^{5,6,11}.

The ages of the patients emerging from a review of the literature varied from 1 to 70 years, while the patient described in this report was 19 years old. The male-to-female ratio in the literature review was 2.5:1⁶. This tumour of the head and neck usually develops as a painless mass with specific symptoms caused by interference with the normal anatomy and physiology of the area in which they are found. The most commonly reported initial symptoms were nasal obstruction, epistaxis, dysphagia and dyspnoea. When the mass is located posteriorly in the oral cavity or tongue, patients may present dysphagia, dyspnoea, snoring, or a combination of these symptoms⁶. Our patient presented a mass on the cheek, associated with swelling, without other symptoms.

Most lesions were treated by local excision without sacrificing structures that would cause major functional or cosmetic morbidity. Our patient was submitted to complete local excision with clear margins without any morbidity. These lesions have no metastatic potential and generally good prognosis. Of the cases with follow-up reported in the literature, only 2 (11%) out of 18 had a recurrence after a local excision^{1,10}. The reason for these recurrences is unknown, as is the adequacy of the margins of resection.

If the surgical margins were reported to be free of tumour, local recurrences were uncommon. Radiation therapy and

chemotherapy have no role in the management of benign FH¹⁶.

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