

Case Report

Dendritic fibromyxolipoma of the parotid gland: a case report and review of the literature

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Abstract: Dendritic fibromyxolipoma, a rare subtype of lipoma, mainly occurs in the neck, back, and chest wall. Here, we describe the first reported case located in the parotid gland. It happened in a 24-year-old woman with a well-demarcated painless mass. Pathologically, the tumor was characterized by small spindle and stellate cells, an unequal number of mature fat cells, plentiful myxoid stroma, and collagenous fiber bundles. Immunohistochemistry demonstrated that the tumor cells were positive for vimentin, CD34, Bcl-2, and desmin, but not for keratin, EMA, SMA, or S-100. The tumor was successfully removed without complications. This report will bring to the attention of clinicians the clinical features and differential diagnosis of this unusual tumor.

Keywords: Dendritic fibromyxolipoma, parotid gland, spindle cell lipoma, solitary fibrous tumor, myxoid liposarcoma

Introduction

Dendritic fibromyxolipoma (DFML) is an uncommon, newly found benign soft tissue tumor that usually presents as a slow-growing, well-defined lesion, most often in adult male patients [1]. Histologically, the characteristic feature is an admixture of spindle cells, stellate cells, mature adipose tissue, and abundant myxoid matrix and obvious collagenization. The tumor is characterized by a positive immune response to vimentin, CD34, and Bcl-2 [1-3]. To date, only 32 cases have been reported and cited in PubMed (**Table 1**), none of which occurred in the parotid gland [1-14]. Here, we report the first case of a 24-year-old woman with DFML in her left parotid gland.

Clinical data

A 24-year-old female presented with a one-year history of a painless, progressively enlarged mass before the left ear. There were no associated constitutional symptoms. The patient had a history of allergy to levofloxacin injection and a family history of hypertension. Physical exam-

ination revealed the lump was about 2.5 cm in diameter, well-defined, soft, inactive, non-tender, and no enlarged lymph nodes were touched in the jaw, behind the ear or in the neck. Computed tomography (CT) demonstrated a hypodense density, a well circumscribed bump in the left parotid gland, measuring 2.1 cm × 1.7 cm of the maximum cross section (**Figure 1**). Well-differentiated adipocytes, spindle cells, and a myxoid background were observed under the microscope of fine needle aspiration cytology (FNAC), which was considered a benign tumor (**Figure 2**). Beyond that, the patient was healthy and all routine laboratory examinations were normal. Then she endured a partial excision and the lump was removed completely.

On gross inspection, the space occupying the lesion was 2.5 cm × 2 cm × 2 cm in size and clearly defined with an intact capsule. The cut surface was grayish white, soft and like a translucent jelly. Under the microscope, the neoplasm was mainly composed of small spindle or stellate cells, variably admixed with mature adipose cells, embedded in a rich mucus, collagen bundles, and blood vessels. At higher magnifi-

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Table 1. Clinical findings of DFML reported in PubMed

Author	Year	Cases	Sex	Age (year)	Tumor size (cm)	Location	Follow-up
Suster [1]	1988	12	11/1 (M/F)	33-81 (median, 64)	2-11 (average, 6)	Neck (4 cases); Back (3 cases); Shoulder (2 cases); Chest wall (2 cases); Face right nasal area (1 case)	5-13 years, NRM
Karim [2]	2003	1	M	73	13	Right shoulder, between infraspinatus and deltoid muscles	8 months, NRM
Tan [3]	2003	8	8/0 (M/F)	45-75 (median, 65)	2-9.5 (average, 4.2)	Neck (2 cases); Shank (2 cases); Left eye socket (1 case); Right shoulder (1 case); Lower back (1 case); Left toe (1 case)	6 months - 3 years, NRM
Al-Maskery [4]	2011	1	F	36	2	Lower lip	2 years, NRM
Dahlin [5]	2012	1	F	65	3.2	Left forearm, adherent to median nerve	NA
Zhang [6]	2013	1	F	32	24	Right inguinal and perineum region	9 months, NRM
Wong [7]	2014	1	M	67	7	Left shoulder	4 months, NRM
Han [8]	2014	1	M	69	1	Nasal tip	NA
Xu [9]	2015	1	M	24	14	Left shoulder, triceps brachii (intramuscular)	4 years, NRM
Liu [10]	2015	1	M	53	2	Right back, latissimus dorsi (intramuscular)	1 year, NRM
Song [11]	2016	1	F	34	4	Right lower jaw	Recurrence after 10 years, no metastasis
Ciloglu [12]	2016	1	F	59	17	Left inguinal region	3 years, NRM
AlAbdulsalam [13]	2016	1	M	38	3.4	Pyiform Sinus	NA
Ruiz [14]	2018	1	M	69	5	Infraclavicular region	2 years, NRM

M, male; F, female; NA, information not available; NRM, no recurrence or metastasis.

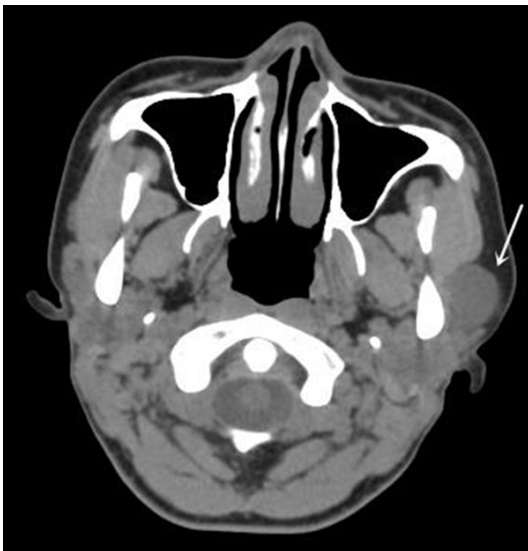


Figure 1. CT scan of the maxillofacial region. A single, boundaries clear protuberance with hypodense density is noted in the left parotid gland (arrow).

cation, the tumor cells were elongated and with dendritic cytoplasmic prominences (**Figure 3**). Immunohistochemical staining showed the spindle and stellate cells stained positive for vimentin (**Figure 4A**), CD34, Bcl-2 (**Figure 4B**)

and desmin, but not for Keratin, EMA, SMA or S-100. Ki-67 showed a low proliferation index < 1%. To date, about 5 months after surgical treatment, the patient is well with no evidence of recurrence or metastasis.

Discussion

As a newly discovered benign tumor, DFML was first described by Suster et al. in 1998, with 12 cases [1]. From then on, about 20 additional cases were reported in 13 articles [2-14]. A review of these cases shows the most common age ranged from 24 to 81, with a median age of 60. The ratio of male to female was about 4.3:1. DFML usually arises in the subcutaneous tissue or superficial fascia of the neck, shoulder, back, and chest wall [1]. Occasionally, it may occur in the intramuscular [2, 9, 10], face [1, 3, 8], shank [3], inguinal and perineum regions [6, 12]. The lesion usually has a complete or partial capsule, with a grayish yellow, gelatinous section, with size varying from 1 to 24 cm (average, 6.3 cm). Microscopically, the neoplasm cells include small spindle cells and astrocytes, which are scattered in a large amount of mucous matrix or collagen fibers. In addition, there are varying amounts of benign

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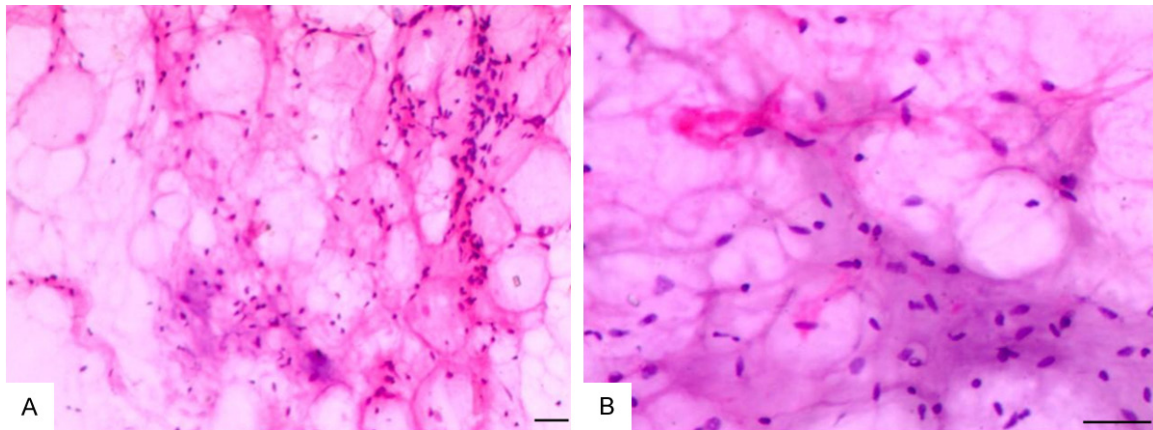


Figure 2. Microscopic results of FNAC. A. Hematoxylin and eosin (HE)-stained section shows the tumor consisted of well-differentiated adipocytes, spindle cells, and myxoid background (HE ×100). B. A higher magnification indicated the nuclei of the tumor cells were round or spindle-shaped without atypia (HE ×200). Bar = 100 μ m.

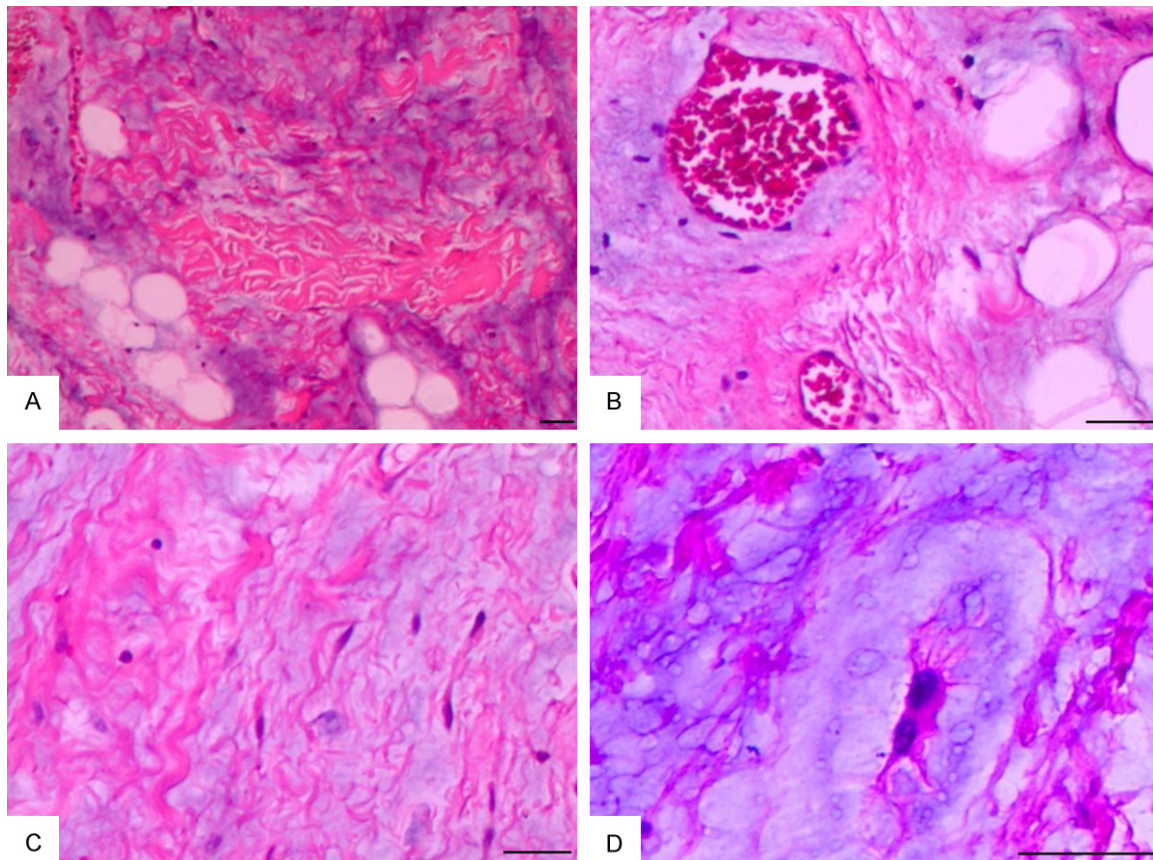


Figure 3. Histopathology of DFML. A. Low-power view showed few tumor cells were dispersed in a matrix of mucus and bulky collagenous fibers, variously mixed with mature lipocytes (HE ×100). B. The stroma was intermixed with thin-walled vessels (HE ×200). C. The elongated small spindle cells (HE ×200). D. Stellate cells with dendritic cytoplasmic prominences (HE ×400). Bar = 100 μ m.

adipose tissue and thin-walled vessels in the background. Immunohistochemical results

show that CD34 and Bcl-2 are expressed in all cases, which could clearly show the dendritic

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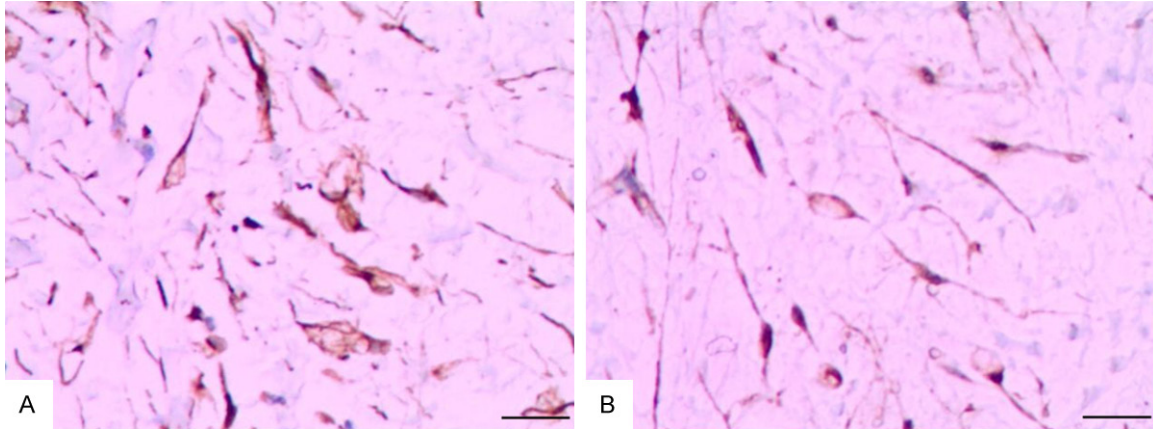


Figure 4. A. An immunohistochemical stain for vimentin showed spindle and stellate cells with many elongated cytoplasmic processes ($\times 200$). B. The proliferating cells stained strongly positive for Bcl-2 ($\times 200$). Bar = 100 μm .

processes of these tumor cells. Our case supported DFML in visual observation, cytology, histology and immunohistochemistry.

Because of the histological characteristics and immunophenotype, DFML is easily confused with other lesions, including spindle cell lipoma (SCL), solitary fibrous tumor (SFT) and myxoid liposarcoma (MLS). SCL is very similar to DFML in terms of age, location, histomorphology and immunophenotype. However, spindle cells in SCL have no obvious dendritic elongated cytoplasmic processes, the interstitium lacks obvious myxoid changes and blood vessels, which are usually thick-walled small blood vessels [1]. At present, the cytogenetic characteristics of DFML have not been clarified. Only one case has demonstrated the presence of the 13q14.3 deletion, which was mostly found in SCL [7]. So some scholars have conjectured that DFML may be a special variant of myxoid SCL [2, 7].

If SFT appears with myxoid stroma or mature adipose tissue, it is easily confused with DFML when the latter is accompanied by more scarring or cord-like collagen fibers with spindle cell proliferation. Generally, SFT shows the alternation of loose and dense areas, but the spindle and stellate cells of DFML are relatively evenly distributed [15]. The spindle shaped cells of SFT are arranged in a fascicular, storiform or hemangiopericytoma-like manner, but they are arranged in a disorderly manner in DFML. Small nucleoli and mitoses can be observed in the tumor cells of SFT, but not in the tumor cells of DFML [15]. Since DFML, SCL and SFT are similar in morphology and immunophenotype,

some experts believe that the three diseases may be the same set of lesions at different stages of maturation, and DFML is a transitional form between SCL and SFT [3]. However, Fritchie et al. [16] showed that there was no correlation between SCL and SFT, since there is a monoallelic or biallelic loss of RB1 by fluorescence in situ hybridization (FISH) in SCL but no loss in SFT. Therefore, the relationship between the three diseases needs further study.

MLS, a low grade malignancy, has similarities with DFML comprising a plentiful myxoid matrix and a plexiform vascular pattern. However, MLS is typically located in the deep soft tissues of the lower limbs, with infiltrating growth, unclear borders, with existing adipoblasts visible on higher magnification, and negative for CD34. In addition, cytogenetic studies had indicated that the molecular feature of MLS was repeated translocations $t(12;16)(q13;p11)$ and, less commonly, $t(12;22)(q13;q12)$, respectively fusing FUS or EWSR1 with DDIT3 on chromosome 12 gene, which could be performed with a DDIT3 separation probe by FISH detection [17, 18].

DFML often presents as a slow growing, well-defined painless mass. The most effective treatment is local excision. Clinical follow-up of the 32 cases ranged from 4 months to 13 years. Only 1 case showed a recurrence after 10 years, but no metastasis [11], and none of the other patients (including our patient) had recurrence or metastasis after their operations. Whether the mechanism of recurrence is related to an incomplete tumor capsule leading

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to postoperative residual remains to be further explored and studied.

DFML in the parotid gland has never been described. Our study reports a case of an unusual location, which was diagnosed comprehensively according to typical clinical characteristics, morphological and immunohistochemical features. It is important for clinicians to recognize the characteristics of this tumor in order to prevent misdiagnosis.

Disclosure of conflict of interest

None.

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