Desmoplastic Fibroblastoma Presenting as a Parotid Tumour: A Case Report and Review of the Literature

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Abstract Desmoplastic fibroblastomas (DFs) are rare fibrous soft tissue tumours that usually arise in subcutaneous tissue or skeletal muscle in a variety of anatomical sites. These lesions most frequently present as painless, slow-growing mobile masses. A case of DF is described in a 47-year-old man who presented with a painless right parotid mass of 2 months duration. At surgery, the lesion was attached to the tail of the right parotid gland. Histopathological examination demonstrated a fibrous lesion comprising spindled and stellate shaped fibroblasts with focal myxoid stromal change. The features were consistent with a DF. This report documents a rare parotid lesion which may mimic other more common parotid gland neoplasms.

Keywords Desmoplastic · Fibroblastoma · Collagenous fibroma

Introduction

The desmoplastic fibroblastoma (DF) (collagenous fibroma) was first described by Evans in 1995 and it was classified as

a distinctive form of benign fibrous soft tissue tumour [1]. These tumours are usually well circumscribed and are composed of spindle to stellate shaped fibroblasts dispersed in a fibromyxoid or densely fibrous background stroma with low mitotic activity. In 1996 the lesion was renamed as a “collagenous fibroma” by Nielsen et al. [2] Since 1995, approximately 94 cases of DF have been reported in the English literature with the largest case series of 63 patients being published by Miettinen and Fetsch [3]. They most frequently present as a non-tender, firm, slow growing mass occurring predominantly in males, with a median age of 50 years. The arm or the shoulders are the most frequent sites of involvement. However, they have also been described in the neck [1–6], tongue [7], lacrimal gland [8] and palate [9, 10]. A rare case of DF presenting as a parotid neoplasm in a 47 year old man is described. To the best of our knowledge, this is the second case report of a lesion presenting in the parotid gland [11].

Case Report

A 47 year old man was referred for investigation and treatment of a painless swelling of the right parotid which was first noticed 2 months previously. There was no history of trauma or infection. On examination, a firm mass was identified which was tucked in below his earlobe and behind the angle of mandible measuring approximately 15 mm maximum dimension. There were no pathological changes of the overlying skin, no regional lymphadenopathy and the patient had normal facial nerve function. On ultrasound examination, a well-defined 11 × 12 × 9 mm mass was identified at the lower pole of the right parotid gland.

A fine needle aspiration biopsy (FNAB) was performed under ultrasound guidance. The FNAB produced tissue

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which showed a spindle cell lesion with features suggestive of nodular fasciitis.

A clinical diagnosis of a benign parotid lesion was made and surgical excision was performed under general anaesthetic. At operation, a firm discrete mass was identified, attached to the tail of the right parotid gland at the anterior border of the sternocleidomastoid muscle. This was resected preserving his facial nerve but sacrificing his greater auricular nerve. Postoperatively, he made an uncomplicated recovery.

Macroscopic examination of the specimen showed a vaguely circumscribed lesion with surrounding glandular parenchyma, the cut surface of which showed an irregular firm white, stellate area measuring $25 \times 15 \times 10$ mm which abutted the surgical margin focally.

Microscopic examination of sections through the tail of the right parotid gland confirmed the presence of part of the major salivary gland which was composed of serous acini with prominent fatty infiltration being noted. The periglandular soft tissues demonstrated a vaguely circumscribed, yet infiltrative spindle cell lesion (Figs. 1, 2). It was relatively paucicellular with a prominent collagenous background stroma (Fig. 3). Interspersed spindled and stellate shaped fibroblasts were observed which had bland nuclei with small inconspicuous nucleoli (Figs. 3, 4). No mitoses were observed. Focally, myxoid stromal changes

were identified within the lesion (Fig. 2). The intervening blood vessels were inconspicuous. There was no evidence of extravasated blood nor chronic inflammation. The lesion was observed extending around and in between skeletal muscle fibres, nerve fibres and into the adjacent parotid gland parenchyma.

Immunohistochemical stains demonstrated focal smooth muscle actin positive staining (Dako, Glostrup, Denmark) while β -catenin showed only weak cytoplasmic staining (Cell Marque, Rocklin California, USA) and was non-specific. S-100 (Novocastra, Newcastle Upon Tyne, UK), and CD34 (Dako, Glostrup, Denmark) were both negative. A diagnosis of DF was made. The patient has been reviewed and 6 months postoperatively is tumour free with no evidence of a recurrence.

Discussion

Desmoplastic fibroblastoma (DF) is a peculiar fibroblastic/myofibroblastic tumour, which has a male predominance and a peak incidence in the fifth and sixth decades of life. DF appears to be more heterogeneous group of tumours than originally envisaged, as evidenced by their presentation in a variety of anatomical sites. In approximately one-third of cases, they characteristically present as slowly enlarging non-tender masses that are noted to increase

Fig. 1 Vaguely circumscribed, spindle cell tumour exhibiting a stellate, infiltrative peripheral margin extending in between parotid gland lobules. (H&E $\times 124$)

Fig. 2 Infiltrative margin of spindle cell tumour extending between skeletal muscle fibres and adipose tissue. (H&E $\times 200$)

Fig. 3 Paucicellular lesion with a prominent collagenous background stroma containing spindled and stellate shaped fibroblasts. (H&E $\times 400$)

slowly in size for more than a year prior to presentation [3]. Sensorimotor neurological deficit is a rare feature and this finding has only previously been described by Fong et al. [4] despite nerve entrapment being a frequent feature [3]. Grossly, collagenous fibromas typically also infiltrate fat and skeletal muscle and this has been observed in 51 % of cases [3]. No mitotic figures or necrotic areas have been documented previously and the blood vessels are usually inconspicuous. Miettinen and Fetsch [3] described two cases in which preceding trauma had been identified in association with the lesions.

The typical histopathologic features of a DF include a paucicellular tumour with uniform stellate or spindle-shaped fibroblastic cells embedded in a highly collagenous or

myxocollagenous stroma. As a result of these findings, Miettinen and Fetsch [3] recommended the designation stellate cell fibroma. Immunohistochemical and ultrastructural studies have shown that the tumour cells are predominantly fibroblastic in nature, although focal myofibroblastic differentiation may be well developed [3]. The tumour cells are typically positive for vimentin and there was often focal reactivity for muscle actins (HHF-35) and alpha-smooth muscle actin [3]. Miettinen and Fetsch [3] have also demonstrated rare keratin-positive cells with the typical stellate and spindle cell morphology [3]. Scattered CD68-positive histiocytes and mast cells may be present, but the tumour cells are negative. There was no documented immunoreactivity for CD34, S-100 protein, desmin, or EMA.

Fig. 4 Collagenous stroma containing spindle and stellate shaped fibroblasts and inconspicuous blood vessels. (H&E $\times 400$)

Table 1 Literature review of desmoplastic fibroblastomas involving the head and neck

Author	Age	Sex	Region	Tumour size	Follow up
Ni et al. [22]	NA	NA	Dorsum nasi	NA	NA
Ahn et al. [8]	53 years	Female	Lacrimal gland	NA	NA
Fong et al. [4]	41 years	Female	Neck	3 cm	No evidence of disease, 18 months
Watanabe et al. [5]	55 years	Male	Neck	3.5 cm	No evidence of disease, 6 months
Bhagalia et al. [23]	58 year s	Female	Oral cavity	2 cm	NA
Shimoyama et al. [10]	49 years	Female	Palate	6 cm	No evidence of disease, 48 months
Ide et al. [11]	50 years	Male	Parotid mass	5 cm	NA
Stacy et al. [24]	40 years	Female	Orbital Rim	NA	NA
Nonaka et al. [7]	87 years	Female	Tongue	0.7 cm	No evidence of disease, 9 months
de Sousa et al. [25]	56 years	Male	Buccal mucosa	0.35 cm	No evidence of disease, 12 months
Evans et al. [1]	58 years	Male	Neck (intramuscular)	1.8 cm	No evidence of disease, 36 months
	25 years	Female	Neck (subcutaneous)	3.2 cm	No evidence of disease, 15 months
Nielsen et al. [2]	47 years	Male	Neck (subcutaneous)	2.2 cm	NA
	76 years	Female	Neck (subcutaneous)	1.5 cm	NA
Hasegawa et al. [6]	37 years	Female	Neck (intramuscular)	5 cm	No evidence of disease, 91 months
Miettinen and Fetsch [3] ^a	NA	NA	Neck	NA	NA
Wilson et al. [26]	88 years	Male	Goiter	4 cm	NA
Mesquita et al. [9]	37 years	Female	Palate	5 cm	No evidence of disease, 6 months

^a Miettinen and Fetsch [3] reported 63 cases of DF of which 14 % were located in the posterior neck or upper back. The tumours ranged from 1.0 to 20.0 cm in maximal dimension (median, 3.0 cm). Follow-up ranging from 46 to 312 months (median, 10.9 years; mean, 12.6 years) was obtained on 39 patients (62 %). No recurrences were documented. However, they did not report data for head and neck DF as a subset

The differential diagnosis included a wide variety of soft tissue lesions including neurofibroma, fibrous histiocytoma, fibromatosis and calcifying fibrous pseudotumour [3, 12]. The principal differential diagnosis in the parotid region would be a solitary fibrous tumour (SFT) [13] desmoid fibromatosis (DFM) [14] or nodular fasciitis.

The SFT is a spindle-cell lesion characterized by a variety of growth patterns within the same lesion classically

described as a ‘patternless pattern’ [13]. All tumours have areas of extensive sclerosis or hyalinization similar to that of a desmoplastic fibroblastoma. SFT could be excluded on the basis of the lack of hypercellular areas, including hemangiopericytoma-like foci which were not identified. Immunohistochemical staining for CD34 helps in distinguishing SFT and DF too, as the present tumour was completely CD34 negative [13].

Desmoid fibromatosis (DFM) presenting as a parotid mass have also been reported [14]. The distinction from DF is difficult, since DF uniformly shows broad areas of marked desmoid-like collagenization. However, the stellate-shaped fibroblasts, a diagnostic trait of DF are always absent in DFM [3]. Unlike DF, DFM is consistently and diffusely more cellular [1–3]. β -Catenin staining is also helpful for discriminating DFM from DF [15, 16] and SFT [17] with positive nuclear reactivity for β -catenin being observed in DFM.

The FNAB result suggested a nodular fasciitis and this is an additional potential differential diagnosis which needed to be considered. Nodular fasciitis is generally easily distinguished from DF. Clinically, it often affects younger individuals and has a rapid onset, often being preceded by a traumatic episode. Histopathologically, nodular fasciitis is generally more cellular and has greater vascularity. Also, it typically has prominent myofibroblastic differentiation with rather strong and diffuse immunoreactivity for muscle actins [18, 19]. Late stages of nodular fasciitis may contain areas reminiscent of those seen in collagenous fibroma, but the former more frequently exhibits cystic degeneration and often contains scattered osteoclast-like giant cells.

Review of the case series and reports published in the literature with involvement of the head and neck indicated that DF occurs primarily in adults. There were ten women and 6 men, ranging in age from 37 to 88 years (mean 53.56 years). The tumours involved the neck, lacrimal gland, palate and oral cavity and they ranged in size from 0.35 to 6 cm (mean 3.08 cm). Follow-up information was available in most cases and ranged from 6 to 91 months (mean 26.77 months). No tumours had recurred or metastasized. (Table 1).

Two translocations t [2, 11] (q31; q12) [20] and t [11, 17] (q12; p11.2) [21] have been detected in DF that had been subjected to cytogenetic analysis. These translocations support a clonal neoplastic process [20, 21].

The recommended treatment of DF is local surgical excision to minimize potential morbidity. There are no reports of local recurrence or metastases to date [1–3, 9, 11, 12].

In conclusion, the present case is the second description of a DF that has involved the parotid gland. The clinical, gross and histologic features are those of a benign neoplasm. Despite only one case having previously been reported in the literature, we highlight this peculiar lesion and wish to increase awareness of these rare lesions amongst surgeons and pathologists alike.

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