Dendritic Myxofibrolipoma: Often Misdiagnosed as Sarcoma

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ABSTRACT

The author describes a benign soft tissue tumor that could be easily mistaken for sarcoma. It represents a combined feature of solitary fibrous tumor and spindle cell lipoma. The clinical presentation, diagnosis, and management of this lesion are discussed. It highlights the importance of proper diagnosis to prevent unnecessary and ineffective treatment by clinicians as the complete excision of this lesion is the treatment of choice and recurrence is very unlikely. It is believed that this case is the first reported case of dendritic myxofibrolipoma occurring in the lower lip mucosa in an Omani patient.

KEYWORDS: Soft tissue neoplasm, lipoma, fibroma, CD34, oral mucosa

Dendritic myxofibrolipoma is a newly described benign soft tissue tumor that could be easily mistaken for sarcoma. It represents a combined feature of solitary fibrous tumor and spindle cell lipoma.¹ It shares a strong positivity for human hematopoietic progenitor cell antigen CD34 and BCL-2 oncoprotein similar to that found in spindle cell lipoma and solitary fibrous tumor.¹⁻⁵ Dendritic myxofibrolipoma is more common in middle-aged and elderly men. It develops primarily in the subcutis or muscular fascia of the head and neck, shoulders, calf, foot, and back.¹ It is well circumscribed macroscopically, and it appears as a partly thin, encapsulated lesion with focal mucinous or gelatinous cutting surface. The tumor is well vascularized histologically and is characterized by an admixture of mature adipose tissue, spindle and stellate cells, and abundant myxoid stroma with prominent collagenization; however, no cytological atypia or mitosis is found. The spindle cells are strongly positive for CD34, BCL-2, and vimentin but negative for S-100 protein and epithelial and muscle markers.⁶ Only 12

cases have been reported in the American and British literature. Few other cases have been reported in the Korean literature and Chinese literature.⁵ To our best knowledge, there have been no case ever been reported in patients from Oman.

CASE REPORT

A 36-year-old Omani woman was referred from a regional hospital with a swelling in the lower lip. She presented to the oral and maxillofacial unit in the Oral Health Department of the Sultan Qaboos University Hospital. The patient had noticed a progressive painless swelling in the mucosa of lower lip that had been slowly increasing in size for the past year. No similar lesions were found elsewhere on the patient's body. There was no history of bursting or bleeding from the lesion. The patient was fit and healthy.

On intraoral examination, there was a 2 × 2-cm round, pink, non-fixed, soft to rubbery soft tissue swelling in the lower lip mucosa, which was very well

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circumscribed with normal overlying mucosa. The patient underwent complete excision of the lesion under local anesthesia. The lesion had a nodular appearance, and it was sent for histopathology examination and immunohistochemical staining. Postoperative healing was uneventful.

FOLLOW-UP AND MANAGEMENT

The excised tumor was sent to the United Kingdom for a second histological opinion. The report confirmed dendritic myxofibroblastoma as the best diagnosis. To prevent local recurrence, complete excision with a normal tissue margin was advised, as the lesion appeared to extend into all margins. The patient underwent a second biopsy as had been advised. The second histological report revealed normal salivary gland tissue, connective tissue, and muscle with foci of foreign body giant cell reaction from previous surgery. The patient was followed for 2 years postoperatively, and there was no evidence of local recurrence or development of a similar lesion.

Macroscopic Examination

The excised lesion was a nodule of soft tissue that was covered by normal mucosal. It measured $2 \times 1.6 \times 2$ cm and the cut surface was shiny with a gray-white area.

Microscopic Examination

The section shows a fairly well-circumscribed lesion with pushing borders as shown in Figs. 1A, 1B and 1C. The lesion comprised loosely arranged spindly or stellate cells in a grossly edematous stroma. The stroma was richly vascular with thin-walled vessels, mainly mucinous, and was focally collagenous. The absence of cellular atypia, necrosis, mitotic activity as well as mature fat cells was evident with scattered mast cells present in the stroma.

Immunohistochemical Examination

The lesion showed a strong immunoreactivity for CD34 in spindle or stellate cells and blood vessels (Fig. 1D). CD99 was strongly positive in spindle or stellate cells. BCL-2 was positive in spindle or stellate cells. CD31 was

Figure 1 (A) Hematoxylin and eosin (H&E)-stained section ($\times 10$) showing abundant myxoid tumor with richly vascular stroma. (B) H&E-stained section ($\times 20$) showing spindle/stellate cells in myxoid stroma. (C) H&E-stained section ($\times 20$) showing collagen fibers in the stroma. (D) Immunohistochemical stain for CD34 shows spindle and stellate cells with many slender dendritic prolongations of the cytoplasm.

positive in blood vessels. Alcian blue 2.5 stroma exhibited a strong positivity. In contrast, immunoreactivity for S-100 protein and desmin was negative. The histopathologic findings revealed a fairly well-circumscribed, richly vascular mesenchymal tumor with a prominently myxoid stroma and with no obvious histological features of malignancy. The lesion was found to be CD34-, CD99-, and BCL-2-positive.

DISCUSSION

Many years have passed since the mesenchymal tumor was first recognized. As a result, it is believed that they are very easy to recognize histopathologically, but pathologists still face challenges in recognizing some of these mesenchymal tumors. The lesion described in this case report is a very distinctive soft tissue lesion that is usually misinterpreted for a low-grade sarcoma (Table 1). This lesion has similarity to myxoid liposarcoma; nevertheless, the absence of atypical lipoblast, the lack of infiltration of the surrounding structure, and being a very well-circumscribed lesion made such diagnosis unlikely.¹ This type of lesion has been reported to occur in patients above the age of 30 years, with a male-to-female ratio of 11:1.1 The tumor usually measures 2 to 11 cm in diameter.¹ Dendritic myxofibrolipoma is mostly reported in the superficial subcutis or muscular fascia, with the most common sites being the head and neck regions, followed by the chest and the back. The results of the

follow-up with this case are in agreement with many studies that reported no local recurrence or distant metastasis of the tumor after simple local excision.^{1,6}

Dendritic fibromyxolipoma is histologically characterized by spindle or stellate fibroblastic cells admixed with adipose tissue in an abundant myxoid and collagenized stroma. The immunohistochemical studies of this type of tumor show strong positivity of tumor cells for CD34, BCL-2, and vimentin. However, it shows negativity for muscle-specific actin (HHF35), desmin, epithelial membrane antigen, Leu 7 (CD57), keratin, and S-100. This tumor appears to represent a transitional form between spindle cell lipoma and solitary fibrous tumor, both of which express strong positivity to CD34 and BCL-2.⁴⁻⁹ Spindle cell lipoma shares a similarity with dendritic myxofibrolipoma. Usually, this type of lesion is well circumscribed, superficially located, and slow growing with few or no lipoblasts. In addition, it exhibits the presence of uniform collagen bundles, which spares vascularity, and an absence of atypical cells.⁴ In contrast, the dendritic myxofibrolipoma is characterized by the dendritic nature of spindle cells, a proliferation of the plexiform capillaries, and the abundance of keloidal-type collagen, which are absent in spindle cell lipoma. The solitary fibrous tumor shares a similarity to spindle cell lipoma, but it differs from dendritic myxofibrolipoma by the lack of mature adipose tissue.⁷⁻⁹ Both spindle cell lipoma and solitary fibrous tumor have been reported to occur in the oral cavity.⁴⁻⁹

Table 1 Characteristic Features Distinguishing Dendritic Myxofibrolipoma and Myxoid Liposarcoma

Features	Dendritic Myxofibrolipoma	Myxoid Liposarcoma
Benign	+	—
Malignant	—	+
Well circumscribed	+	+/-
Encapsulated	+	Partially
Superficially located	+	—
Prominent vascularity	+	+
Invasion/infiltration	—	+
Dendritic nature of spindle cells	+	—
Prominent myxoid and collagenized stroma	+	—
Mature normal looking adipose tissue	+	—
CD34 +	+	—
BCL-2 +	+	—
S-100 +	—	+
Well-differentiated lipoblast	—	+
Mitotic figure/necrosis/atopia	—	+
Lipoblast	—	+
Atypical adipocytes	—	+
Mature adipose tissue	+	+
Metastasis	—	+
Local recurrence	—	+

+, feature present; —, feature absent.

The lesion removed from the patient was a well-circumscribed, nodular lesion located in the mucosa of the lower lip. This lesion was CD34- and BCL-2-positive, like spindle cell lipoma and solitary fibrous tumor. Nonetheless, all reported cases show a mixture of mature fat cells, which were absent in the lesion removed from our patient. As a result, the case was sent abroad for expert opinion, which confirmed the diagnosis of dendritic myxofibrolipoma. Complete surgical excision appears to be the treatment of choice, which was performed on this patient.

CONCLUSION

This article describes a rare lesion occurring in the oral cavity of a 36-year-old woman. It brings the attention of clinicians to the importance of proper identification and characterization of this tumor. This will aid clinicians to avoid misdiagnosis and confusion with other, more aggressive conditions occurring in the head and neck region, such as myxoid liposarcoma and myxoid malignant fibrous histiocytoma. The treatment of this case is remedied by simple excision of the lesion and a close follow-up. This is the first case of a rare dendritic myxofibrolipoma reported in Oman.

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