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CASE REPORT

Pleomorphic hyalinizing angiectactic tumor of the buccal mucosa

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Pleomorphic hyalinizing angiectactic tumor (PHAT) of soft parts is a recently defined mesenchymal tumor of uncertain differentiation, with only a limited number of examples reported to date. We present a case of PHAT of the buccal mucosa in an 86-year-old woman. To our knowledge, there is no formal description of this tumor in the oral cavity.
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An 86-year-old woman presented with a painless, slow-growing lump in her buccal mucosa, which had been present for 6 months. Examination confirmed the presence of a circumscribed, non-tender mass of the right buccal mucosa, partially fixed to the underlying muscle (Fig. 1). Two years follow-up showed no signs of recurrence. The excised specimen, measuring $3 \times 3 \times 2$ cm, was a well-demarcated but non-encapsulated fibrous mass with numerous ectactic vessels (Fig. 2a). Microscopic examination revealed a variegated tumor composed of hyalinizing angiomatous foci and cellular components (Fig. 2b). Cellularity varied from area to area but tumor cells tended to be located near the clustered, angiectactic vessels with perivascular hyalinization (Fig. 2c). They showed considerable degenerative nuclear atypia such as pleomorphism and hyperchromatism and often contained intranuclear cytoplasmic inclusions (Fig. 2c,d). Mitotic figures were rarely observed. Hypocellular areas were characterized by hyalinized or myxoid stroma. Although typical Antoni A and B type structures could not be found, a diagnosis of ancient schwannoma was provided before immunohistochemical examination. Plump, spindled and rounded tumor cells were diffusely and intensely immunopositive for vimentin (V9, 1:200; Dako, Glostrup, Denmark) and CD34 [MY10, 1:50; Becton Dickinson, San Jose, CA, USA, (Fig. 2e)] but uniformly negative for S-100 protein (polyclonal, 1:1000; Dako), α-smooth muscle actin (1A4, 1:800; Dako), desmin (D33, 1:200; Dako), CD31 (JC70, 1:80; Dako), and CD68 (Kip-1, 1:200; Dako). The final diagnosis rendered was pleomorphic hyalinizing angiectactic tumor.

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Comments

In 1996, Smith et al. (1) first described 14 cases of an unusual mesenchymal tumor composed of combination of sheets of pleomorphic cells associated with the hyalinized angiectactic vasculature under the more descriptive term *pleomorphic hyalinizing angiectactic tumor* (PHAT) of soft parts. Since then, only six additional cases have been reported in published work (2, 3). It occurs mostly in the superficial subcutaneous tissue of the lower extremities of adults. Obviously, PHAT acts in a benign fashion but has the potential for local recurrence. It still needs to be clarified whether PHAT may be regarded as a low-grade sarcoma (2, 3).

The PHAT shares several morphological features with schwannoma and malignant fibrous histiocytoma (1, 4). Especially, it shows a striking resemblance to an ancient variant of schwannoma. Marked nuclear pleomorphism and hyperchromatism, intranuclear cytoplasmic pseudoinclusion formation, remarkably prominent vascular changes and widespread stromal hyalinization may not be specific for PHAT (1, 4, 5); therefore, strict criteria differentiating between PHAT and ancient schwannoma are difficult to establish (5). The only clue to the differential diagnosis was the absence of fibrous capsule in PHAT (1, 4). The present case encountered nearly 20 years ago was mistakenly regarded as schwannoma with advanced degenerative changes until it was re-examined immunohistochemically. The diagnosis of PHAT in our tumor was supported by the absence of S-100 protein and the presence of CD34 in tumor cells, thus indicating the possibility that some older cases of ancient schwannoma defined prior to the advent of immunohistochemistry may

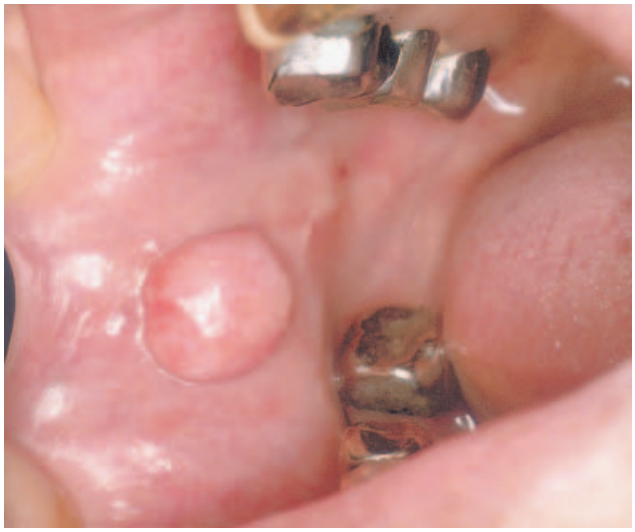

Figure 1 Dome-shaped mass of the buccal mucosa.

be reclassified as PHAT. Recently, the expression of CD99 and vascular endothelial growth factor has been observed in PHAT (2) but the diagnostic specificity remains speculative.

Despite the immunohistochemical and ultrastructural analyses (1–3), the constituent cells of PHAT do not show any features of a specific mesenchymal lineage (4, 5); therefore, some authors consider that PHAT may be the degenerated form of neural, fibrous and vascular tumors (5). Given the overlapping histologic feature and substantial expression of CD34, PHAT is histogenetically akin to the other CD34-positive mesenchymal tumors such as solitary fibrous tumor and giant cell angiofibroma (2, 3).

In summary, we describe the first example of PHAT in the oral cavity. The lesson to be learnt from this case is the necessity for complementary immunohistochemistry for unqualified oral tumors of mesenchymal origin.

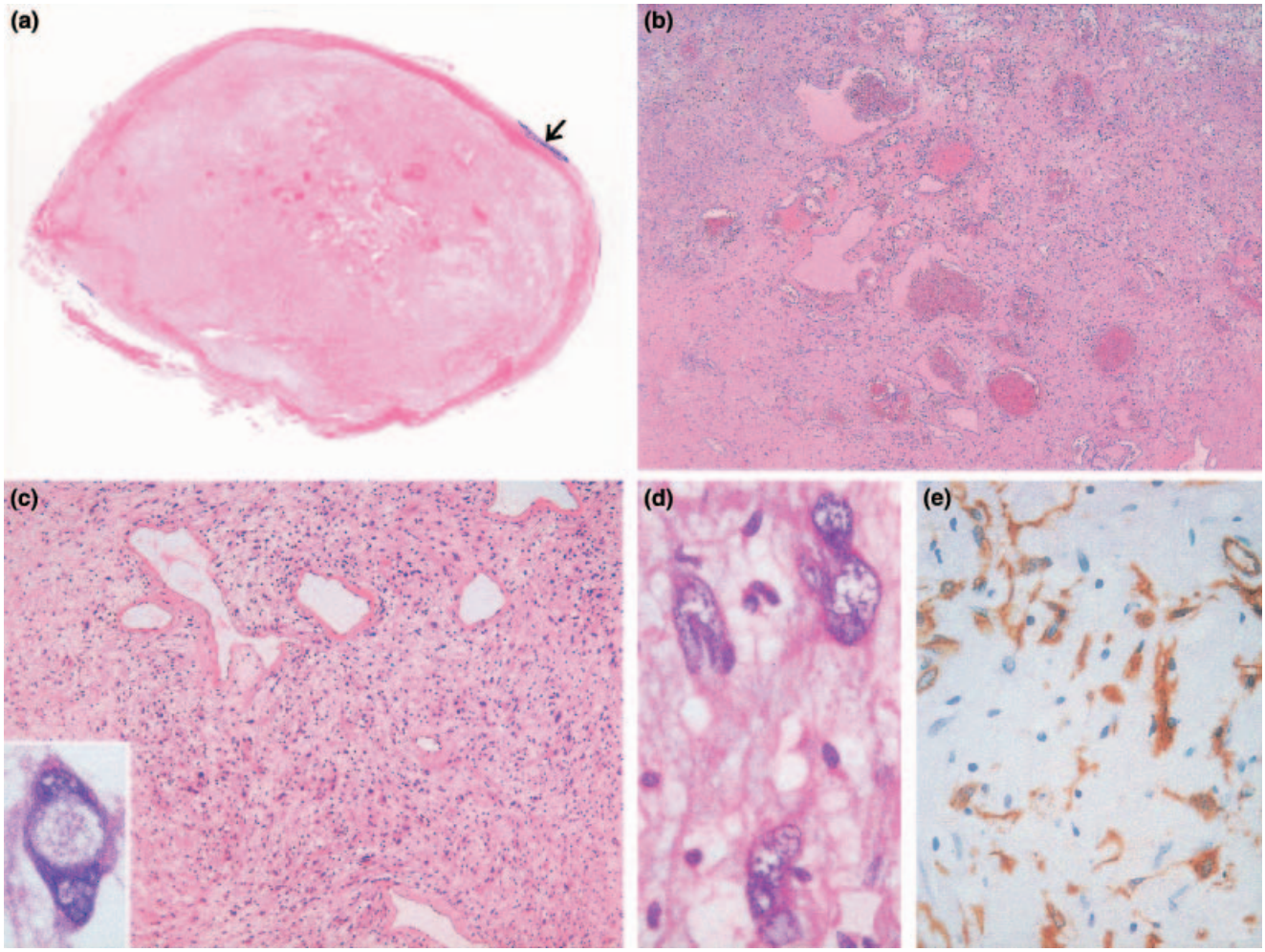

Figure 2 (a) Circumscribed but unencapsulated angiomatous tumor. Compressed lymphoid tissue is indicated by the arrow. (b) Numerous thin-walled ectactic vessels arranged in clusters. (c) Plump spindled to rounded tumor cells with pleomorphic nuclei around the hyalinized angiectactic vessels. *Inset*: Intranuclear cytoplasmic inclusion. (d) Pleomorphic tumor cells. (Hematoxylin and eosin stain, a, ×3, b, ×40, c, ×100, *Inset* and d, ×400). (e) Clear-cut CD34 immunopositivity in tumor cells (avidin-biotin-peroxidase complex method, ×200).

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