A Myopericytoma of the Nose

Ikuko Numata, Satoshi Nakagawa, Satoshi Hasegawa and Setsuya Aiba

Department of Dermatology, Tohoku University Graduate School of Medicine, 1-1 Seiryo-machi, Aobaku, Sendai 980-8574, Japan. E-mail: ikk@mail. tains.tohoku.ac.jp

Accepted October 14, 2009.

Sir,

Myopericytoma (MPC) is a benign mesenchymal tumour composed of oval- to spindle-shaped myoid-appearing cells with a striking tendency for concentric perivascular growth. The vast majority of cases arise in the dermis and subcutaneous tissue, with an apparent predilection for the extremities. The classical presentation is a slowgrowing painless nodule, although occasional cases are painful. We describe here a case with a large tumour on the nose, approximately 4 cm in diameter, which was histologically diagnosed as MPC.

CASE REPORT

A 59-year-old Japanese man presented with a history of a painless mass on the left wing of his nose, which had developed over 5 years. The mass had increased in size gradually. Physical examination revealed a soft, elastic, skin-coloured tumour with telangiectasia, $45 \times 40 \times 36$ mm in size (Fig. 1). He had no history of injury to the nose. He had been treated previously for hypertension, asthma, gastric ulcer and intestinal infarction, but his current general health was good, and he was not taking any medication. There were no abnormal findings in routine laboratory examinations, and no family history of similar cutaneous tumours.

Ultrasound examination revealed a low echogenic mass with lower reflective areas inside it and with marked internal vascularity on Doppler examination.



Fig. 1. A soft, elastic, skin-coloured lesion on the dorsal nose.

Computed tomography (CT) showed an exophytic round mass lesion, 3.6 cm in size, of mainly middle density, although there were several small hyperdense foci, which suggested areas of calcification and a few hypodense areas. Use of an iodine-based contrast medium during CT showed a centripetal and progressive enhancement on early to delayed phase. Magnetic resonance imaging of the tumour showed predominantly a signal isointense to muscle with small hyperintense foci within it on T1-weighted images. On T2-weighted images, it was mostly hyperintense with hypointense foci. Intravenous administration of gadolinium demonstrated intense enhancement of the majority of the tumour except for a small hyperintense focus on unenhanced images, which suggested an area of haemorrhage or calcification within the lesion. No arteriovenous shunt was revealed on magnetic resonance digital subtraction angiography.

Histological assessment of a punch biopsy demonstrated an unencapsulated neoplasm situated in the deep dermis with extension into the subcutis (Fig. 2a). There was a proliferation of bland round to ovoid cells with the eosinophilic cytoplasm arranged in a concentric perivascular pattern and many thin-walled branching staghorn vessels (Fig. 2b). Immunohistochemical studies revealed that the tumour cells were positive for vimentin, α -SMA, and muscle-specific actin (HHF35), but negative for CD34 and desmin. No mitotic figure was observed and less than 3% of cells stained positively for Ki-67.

From the clinical and histological findings we made a diagnosis of MPC. The patient did not agree to undergo surgical resection, and decided to leave the lesions untreated. His tumour had not increased in size at a 4-month follow-up examination and he refused further follow-up.

DISCUSSION

MPC is a relatively new disease entity, described by Granter et al. in 1998 (1). The lesion typically arises within the subcutaneous tissue of the extremities in adults. Histologically, it is characterized by perivascular proliferation of oval- to spindle-shaped cells exhibiting differentiation toward pericytes (2, 3). This arrangement may have a multilayered and concentric appearance. It presents as a well-circumscribed, unencapsulated tumour.





Fig. 2. (a) Unencapsulated neoplasm situated in the deep dermis with extension into the subcutis (*arrows*). (b) A proliferation of bland round to ovoid cells arranged in a concentric perivascular pattern and many thin-walled branching staghorn vessels (haematoxylin and eosin staining).

MPC presents as a benign, slow-growing nodule, and may occasionally be painful. Tumours rarely exceed 2 cm in size. A rare malignant transformation histologically characterized by high cellularity, significant mitotic activity, pleomorphism, and necrosis has also been reported (4).

MPC shares morphological features with solitary fibrous tumours, glomus tumours, and myofibromata (2) and should be differentiated from these tumours. Application of strict morphological criteria and appropriately selective immunohistochemical markers will help in distinguishing MPC from its mimics. Immunohistochemically, the tumour cells express positive reactivity for α -SMA (5), muscle-specific actin (3, 5) and vimentin (3), and are negative for desmin (5) and CD34 (6).

No cases of this rare mesenchymal neoplasm located on the nose have been reported previously. We should keep in mind that a nodule of the nasal region may be a myopericytoma.

REFERENCES

- Granter SR, Badizadegan K, Fletcher CD. Myofibromatosis in adults, glomangiopericytoma, and myopericytoma: a spectrum of tumors showing perivascular myoid differentiation. Am J Surg Pathol 1998; 22: 513–525.
- Datta V, Rawal YB, Mincer HH, Anderson MK. Myopericytoma of the oral cavity. Head Neck 2007; 29: 605–608.
- Sadahira C, Yoneda K, Moriue T, Takai I, Kushida Y, Haba R, et al. Periungual myopericytoma. J Am Acad Dermatol 2006; 54: 1107–1108.
- McMenamin ME, Fletcher CD. Malignant myopericytoma: expanding the spectrum of tumours with myopericytic differentiation. Histopathology 2002; 41: 450–460.
- 5. Matsuyama A, Hisaoka M, Hashimoto H. Angioleiomyoma: a clinicopathologic and immunohistochemical reappraisal with special reference to the correlation with myopericytoma. Hum Pathol 2007; 38: 645–651.
- Dray MS, McCarthy SW, Palmer AA, Bonar SF, Stalley PD, Marjoniemi V, et al. Myopericytoma: a unifying term for a spectrum of tumours that show overlapping features with myofibroma. A review of 14 cases. J Clin Pathol 2006; 59: 67–73.