ANGIOSARCOMA OF THE PAROTID GLAND

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Summary

Angiosarcomas of the major salivary glands are rare tumours. The authors describe a case of the tumour located in the right parotid gland of a 77-year-old woman. Histological examination revealed a poorly differentiated tumour made up of epithelioid and spindle cells. These two types of cells intermingled. In some parts, primitive mutually anastomosing irregularly shaped vascular spaces with atypical endothelial cells were found. The tumour cells were positive for CD31, CD34, EMA and FVIII (focally). Due to the relatively short follow-up period the prognosis of the disease is difficult to estimate.

Key words: angiosarcoma – parotid gland – immunohistochemistry

Souhrn

Angiosarkom příušní žlázy

Angiosarkom velkých slinných žláz patří ke vzácně se vyskytujícím nádorům. Autoři uvádějí jeden takový nádor lokalizovaný v pravé příušní žláze 77leté ženy. Histologické vyšetření ukázalo, že šlo o nediferencovaný nádor tvořený epiteloidními strukturami a protáhlými nádorovými buňkami. Oba buněčné typy v sebe vzájemně přecházejí. Místy byly zjištěny i primitivní, vzájemně anastomozující nepravidelné cévní prostory s atypickými endoteliemi. Nádor pozitivně reagoval s protilátkami proti CD31, CD34, EMA a FVIII (ložiskovitě) antigenům. K prognóze onemocnění je zatím obtížné se vyslovit vzhledem ke klinicky relativně krátké době sledování pacientky.

Klíčová slova: angiosarkom – parotis – imunohistologie

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Angiosarcomas of the major salivary glands are rare. They were described either as isolated or together with the same tumours in the oral region (2, 4, 6, 7, 10, 11). Besides primary angiosarcomas of the salivary glands, secondary tumours were observed. Fanburg-Smith et al. (3) found that angiosarcomas of the oral cavity and salivary glands included 7 secondary angiosarcomas of which 3 involved the parotid gland. In some cases, angiosarcoma is linked to a benign vascular tumour such as congenital haemangioma (2).

CASE REPORT

A 77-year-old woman presented with an enlarged right parotid gland. Clinical examination revealed a tumour which was surgically removed. Macroscopically, the mass was relatively well-defined, solid, grey-white, sized 2 x 1.5 x 2 cm. The patient was treated with radiotherapy. One year postoperatively, the patient was without clinical signs of recurrence. Detailed clinical examination did not reveal any other primary tumour.

Histologically, the tumour was made up mostly of spindle cells, with a gradual transition into areas composed of large epithelioid cells (Figs. 1, 2). These cells had abundant, eosin-stained cytoplasm, vesicular nuclei and distinct nucleoli. Some areas were less cellular, made of connective tissue, with irregular narrow, mutually anastomosing vascular spaces. These spaces were lined by atypical endothelial cells (Fig. 3). The tumour contained necrotic areas. Numerous mitoses were noted.

For immunohistochemistry paraffin embedded tissues were examined. Sections were deparaffinized and the following primary antibodies were used (dilution in parenthesis): CD34 – clone QBEnd-10 (1:50), von Willebrand factor (factor VIII; 1:100), vimentin – clone Vim3B4 (1:50), AE1-AE3 (1:50), S-100 protein (1:100), SMA – clone 1A4 (smooth muscle actin; 1:50), Ki-67 – clone MIB-1 (1:100) all from DAKO, Glostrup; CD31 (1:40), EMA – clone GP1.4 (epithelial membrane antigen; 1:50), produced by Novocastra, Newcastle. We used an avidin-biotin conjugate (ABC) method. The colour was developed with diaminobenzidine, supplemented with hydrogen peroxide. Immunohistological examination showed positive results with antibodies against vimentin, CD31, CD34, epithelial membrane antigen (EMA) (Figs. 4-6) and von Willebrand factor (factor VIII). Negative results were obtained in tests with antibodies against cytokeratins AE1/AE3, S-100 protein and smooth muscle actin (SMA). The proliferation marker Ki-67 was positive in "hot spots" of 60-80 % of cells.

DISCUSSION

Malignant mesenchymal tumours of the major salivary glands are uncommon findings. As of 1986, sixty-seven sarcomas or sarcomatoid lesions were recorded in the AFIP registry of salivary glands, including 4 primary angiosarcomas or malignant haemangioendotheliomas, 11 malignant schwannomas, 9 fibrosarcomas and 4 malignant fibrous histiocytomas (1). Most frequently, the parotid glands are affected; angiosarcomas of the submandibular glands are rare (3, 10, 11). According to some authors (3), angiosarcomas localized in the oral cavity and major salivary glands account for 2% of all angiosarcomas. Most frequently, they develop in