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ADENOID CYSTIC CARCINOMA OF THE SKIN

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Adenoid cystic carcinoma commonly arises from the major and minor salivary glands. Primary cutaneous adenoid cystic carcinoma is extremely rare. It was seen in a 75-year-old male as a large tumour over the sternal and pectoral region. The tumour was hard, non-tender, and fixed to the underlying structures with visible congested veins. Histopathologically, large cell masses with adenoid or cribriform pattern, forming cystic spaces at places were seen. Solid epithelial nests and a few ductal structures were also seen. Pulmonary metastases were another unusual feature in our patient perhaps not reported before.

Key words : Adenoid cystic carcinoma.

Adenoid cystic carcinoma commonly arises in the salivary glands. Originally called cylindroma by Billroth, this generic name was used for almost 100 years till the specific name of adenoid cystic carcinoma was first used by Spies in 1930[1] and later popularised by Foote and Frazell in 1953.[2] Besides salivary glands, adenoid cystic carcinomas have also been reported to occur in lacrimal glands, ceruminous glands of external auditory canal, mucosal glands of upper airway, oesophagus, breast, Bartholin's glands, uterine cervix and prostate.[3] Primary adenoid cystic carcinoma of the skin is an exceedingly rare entity. Cooper reported a solitary case and could find only 7 other cases from the available literature till 1984.[4] The rarity of the tumour and its aggressive clinical behaviour prompted us to report the case.

Case Report

A 75-year-old male patient presented in March 1986 with a recurrent, painless swelling in front of the chest of six months duration. A lump had been excised from the same site at a district hospital six months ago. However, the details of the operation and histopathological report were not available. The patient was found to have a 25×25 cm size swelling over the sternal and left pectoral regions of anterior chest wall with two prominant humps and a well-healed scar of previous excision (Fig. 1). Congested veins were found over the tumour which was hard, non-tender and fixed to the overlying skin and underlying pectoralis major and sternum. There was no axillary or inguinal lymphadenopathy. Systemic examination was non-contributory.

Routine blood counts, blood urea, sugar and LFT were normal. Skiagram chest revealed two, large, extra-thoracic soft tissue shadows without any calcification. There were however,

Fig. 1. Large dumb-bell, shaped tumour in the anterior chest wall.

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multiple, bilateral, rounded opacities in the lungs suggesting pulmonary metastases. As a palliative measure, a wide excision of the tumour including underlying pectoralis major muscle and anterior cortex of sternum was done with primary closure. The tumour was very vascular and excision required 3 units of blood transfusion. Post-operative recovery was marred by an area of flap necrosis which slowly healed over the next 4 weeks.

Naked eye examination of the specimen showed a dumb-bell shaped swelling consisting of two globular masses connected together by an isthmus covered by skin measuring $20 \times 9 \times 7$ cm. Cut section revealed a fleshy and lobular pattern. The tumour appeared well encapsulated. Multiple sections from various representative areas revealed large cell masses with an adenoid or cribriform pattern, at places forming cystic spaces lined by flattened cuboidal epithelium. Solid epithelial nests and a few ductal structures were also seen in some areas. The overlying skin was free and there was no continuity of the tumour with it. Invasion of the perineural spaces was seen in many areas (Fig. 2).

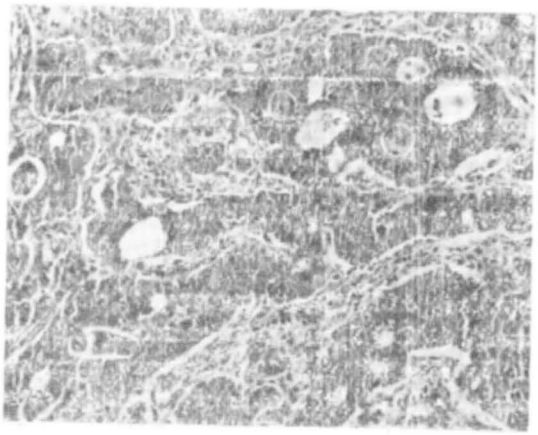

Fig. 2. Adenoid cystic pattern of the tumour (H and E X 150).

Comments

Adenoid cystic carcinoma arises most commonly in the major and minor salivary glands.[3] Cutaneous involvement can result from perineural or direct extension of the salivary gland neoplasm.[5] Cutaneous adenoid cystic carcinoma in a location remote from adjacent salivary tissue is rare.[6] From the available literature this is the ninth well-documented example of primary cutaneous adenoid cystic carcinoma excluding lesions of external auditory canal. Marsh and Allen[7] studied 38 cases over a period of 32 years, but could not find any cutaneous involvement. Similarly, Smith et al[8] in a series of 58 cases of ACC did not find any tumour in the skin. Cutaneous adenocystic carcinoma has been found to affect patients of middle and older age groups with a predilection for women.[3] The duration of the tumour is known to vary from 6 months to 14 years. The common sites of involvement reported so far are scalp, arm, abdomen, axilla and nose.[4] The tumour usually appears as discrete nodules but confluent nodular masses or diffuse thickening of the dermis have also been reported. Pain is not a constant feature. Loss of hair from skin over the tumour has been reported.[3] The maximum size of tumour has been reported to be 8 cm by Cooper[4] but in the present case the diameter of the tumour measured 20 cm.

Adenoid cystic carcinoma usually carries a predictable natural history. Indolent, yet locally aggressive, it can infiltrate into the surrounding tissues especially in the perineural planes. Adherence to the brachial plexus and skeletal muscles have been reported[3] but infiltration of bone or other anatomical structures has not been described so far.[4]

Microscopically, the tumour resembles ACC elsewhere, but in the skin it must be differentiated from an aggressive basal cell carcinoma. This differentiation is based on the combined obser-

vation of, lack of continuity with the epidermis or hair sheath, some membrane-bound tumour lobules without palisading, and very prominent perineural infiltration.

The tumour is well known for local recurrence, hence wide surgical excision with a meticulous histological control of the margins is the only appropriate treatment. Radiotherapy is of doubtful value. Though regional lymph nodes may be invaded by contiguity, true embolic lymph node metastasis is rare.[9] In cutaneous ACC, lymphatic metastases are unknown, though lymph nodal dissection has been carried out in 2 patients.[3,4] Distant metastases occur in 40-50% cases of non-cutaneous ACC late in the course of disease, with lungs, bones and brain being the common sites.[7] In ACC of the skin, distant metastases have not yet been reported.[4] Our case represents perhaps the first such report of pulmonary metastases from a cutaneous ACC.

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