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PRIMARY MUCINOUS CARCINOMA OF SKIN

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A 65-year-old male had recurrent primary mucinous carcinoma of skin arising in the upper one third of the left forearm. It was characterized by the presence of a large area of mucin with the tumour cells present in sheets and arranged in duct-like structures. Primary mucinous carcinoma is characterized by a very low incidence of metastases. The mucinous material seen in the primary mucinous carcinoma is non-reactive to alcian blue and aldehyde fuchsin at a very low pH (0.4).

Key words : Mucinous carcinoma, Skin.

Primary mucinous carcinoma of the skin is an extremely uncommon skin tumour. Lennox et al[1] reported the first case of mucoid carcinoma on the scalp. Mendoza and Helwig[2] reported 14 cases and discussed the clinical and histochemical characteristics of these tumours in detail. Rodrigues et al,[3] Grossman and Izuno[4] and Metz et al[5] reported single cases. Headington[6] reviewed all the 20 cases reported in the literature and added 2 of his own. Of the 22 cases reported, 15 were located on the head and neck. Metastases were present in only 3 cases involving regional lymph nodes. The present case was located at an unusual site.

Case Report

A 65-year-old male patient was admitted with a gradually increasing, painful and ulcerated swelling in the left forearm just below the elbow for the last 6 months. Initially, a swelling was noticed 10 years back which gradually increased in size without any pain or ulceration. There was no history of trauma. First local excision was done 6 years back. Physical examination revealed a soft to firm mass, 6×4 cm in size, with irregular everted margins and serosanguinous discharge (Fig. 1). The tumour mass was freely mobile in either direction. The regional or distant lymph nodes were not palpable. X-ray of the chest and elbow joint did not reveal any abnormal features. Other laboratory investigations were normal. The tumour mass was excised with surrounding normal skin tissue.

Fig. 1. A round, elevated, sharply demarcated ulcerated mass just below the elbow.

Grossly, the tumour mass measured 5×4×2.6 cm in size with an ulcerated area of 1.5×1.4 cm in size with healthy skin all around. On cut section, the tumour mass was sharply demarcated underneath the skin, with a honeycomb appearance due to gelatinous areas at places.

Microscopically, sections stained with haematoxylin and eosin revealed a circumscribed lesion in the dermis composed of nests of

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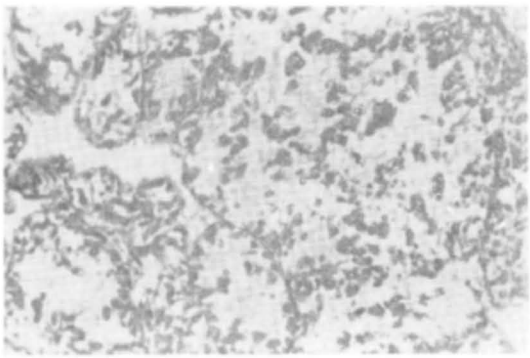

Fig. 2. Nests of epithelial cells exhibiting solid and glandular patterns with large mucinous pools separated by thin fibrous septa (H and E×100).

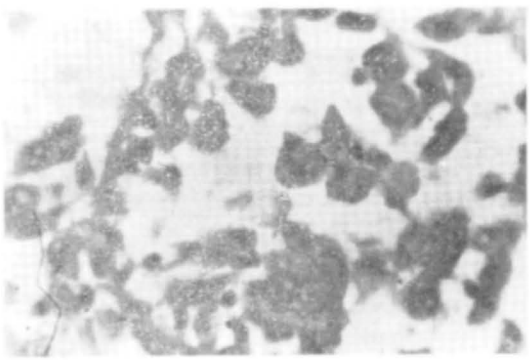

Fig. 3. Groups of malignant cells, having a moderate amount of cytoplasm and hyperchromatic nuclei with prominent nucleoli, lying in a mucinous pool (H and E×450).

solid and glandular epithelium with a pool of mucin (Fig. 2). The epithelial cells showed eosinophilic cytoplasm with round hyperchromatic nuclei (Fig. 3). These cells were seen lying scattered or in groups in the mucinous pool, at places separated by thin fibrous septa.

Paraffin sections stained with PAS and DPAS revealed diastase resistant PAS positive material. The mucinous material also reacted positively with mucicarmine, alcian blue (pH 2.5) and aldehyde fuchsin (pH 1.7). The mucinous material was non-reactive at very low pH (0.4) with alcian blue and aldehyde fuchsin. A diagnosis of primary mucinous carcinoma of the skin was made.

The swelling recurred twice at the same site 4 years after the first excision and 3 months after the second excision. On the third admission, the limb was amputated 15 cm above the elbow joint. Histopathological examination of the material was again suggestive of mucinous carcinoma of skin.

Comments

Mucinous carcinoma is probably derived from sweat glands or from the derivatives of the primary germs of these glands. Histopathologically, it is characterised by the presence of a large pool of mucin with tumour cells arranged in sheets, clusters or duct-like structures. Histochemically, it contains a non-sulphated mucoprotein, probably sialomucin.[2] The histopathological distinction between the sweat gland carcinoma and primary mucinous carcinoma is less clear. The presence of a large pool of mucin, non-reactive to alcian blue and aldehyde fuchsin at low pH (0.4) is always indicative of primary mucinous carcinoma of skin, whereas mucin present in the sweat gland carcinoma is strongly reactive to these stains at the low pH (0.4) and is resistant to sialidase treatment.[2]

Of the 22 cases reviewed by Headington,[6] 15 cases were seen on the head and neck region, 3 cases in the axilla, 2 cases involved the thorax and 1 case each the abdomen and foot. The age of the patients ranged from 8 years to 76 years. Duration ranged from 1 month to 5 years. Seventeen cases were males and 5 cases females. Local excision was done in all the cases except one case in which a wide excision was followed by radiotherapy.[5] Recurrence was seen in seven cases. These tumours tend to ulcerate and recur frequently.[2,7] The present case also showed recurrence. The tumour has a characteristic locally aggressive behaviour and in most cases local excision is the treatment of choice,

which may be responsible for high recurrence rate. The low incidence of metastases in the primary mucinous carcinoma of skin in contrast to the sweat gland carcinoma may probably be related to the mucin-secreting ability of the tumour. Norris and Taylor[8] suggested that some carcinomas of the breast have a better prognosis by virtue of the marked mucin secretion than those with low mucin secretion. Mendoza and Helwig[2] suggested that the property to secrete mucin is an uncontrolled function of these tumours and may be evidence of a high degree of dedifferentiation. This accumulation of mucin may interfere with the nutrition of the tumour cells due to its increased pressure effect, which in turn could affect the rate of cell reproduction and correlate with the relatively benign behaviour of the tumour.[2] Up till now distant metastases has not been reported, though regional lymph nodes were affected in 3 patients.[6]

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