Squamous cell carcinoma arising in a subcutaneous dermoid cyst

G.C. Stephenson and J.W. Ironside

Departments of Neurosurgery and Pathology, The General Infirmary at Leeds, Leeds LS1 3EX, UK.

Summary: A case of malignant transformation to a squamous cell carcinoma in a long-standing subcutaneous dermoid cyst in a 44 year old man is presented. Malignant transformation in dermoid cysts is extremely uncommon and has never previously been recorded at this site. The need for adequate investigation and treatment of superficial midline lesions presenting in childhood is emphasized.

Introduction

Superficial dermoid cysts are histologically benign maldevelopmental tumours usually evident at birth. These tumours enlarge by desquamation of keratin and lining cells into a central cystic cavity and by the growth of skin adnexal components such as hair follicles. Malignant transformation in a dermoid, although exceptional, has previously been described in ovarian and intracerebral sites, but not in a lumbosacral subcutaneous cyst.

Case report

A 44 year old typewriter engineer was first seen in a neurosurgical outpatient clinic in August 1988 complaining of an uncomfortable enlarging swelling over his lower back. He was born with a swelling over his lumbosacral region with an overlying hairy patch and was originally thought to be suffering from spina bifida occulta. This suggestion was not substantiated by later spinal radiographs. He first sought medical attention in 1972 because of the increasing size of the tumour, but was told that excision would require major surgery and the risks outweighed the benefits.

Examination revealed a healthy looking man with no neurological signs. A large cystic swelling occupied most of the lower back (Figure 1). A myelogram and a computerized tomographic (CT) scan revealed the tumour to be of mixed density and entirely subcutaneous. In particular no communication with the spinal cord was established. It was felt that the findings were consistent with a dermoid cyst. Other routine investigations were normal.

The tumour (25 × 23 × 20 cm) was subsequently removed with an ellipse of overlying skin (24 × 7 cm). At operation the tumour was found to consist mainly of pultaceous material, but part of the cyst wall was very adherent to the paravertebral tissues in the region of the 4th lumbar vertebra and looked frankly neoplastic. On opening, the cystic component of the tumour appeared multiloculated with a wall measuring up to 5 mm in thickness. The cyst contents included hair, keratinous debris and oily material (Figure 2). Histological examination of sections from the thinner part of the cyst wall showed a lining of keratinized squamous epithelium and evidence of a foreign body giant cell reaction to released keratin. The thicker areas of the cyst wall showed widespread sheets and anasto-
extended into the ligamentous and cartilagenous paravertebral tissues.

Post operatively he was referred for radiotherapy, but a further CT scan performed 4 weeks after the operation revealed a massive recurrence of the malignant component of the tumour. This was re-explored and the tumour was found to be extending through the paravertebral muscles to the laminae of L4 but not invading the dura. This was excised and histology confirmed a squamous cell carcinoma.

The patient represented 2 weeks later with further subcutaneous recurrences widely disseminated in the entirety of the explored area. It became evident at this stage that further surgery would not be beneficial and local radiotherapy was embarked upon. He has been reviewed since and although he remains neurologically intact his tumour has not responded well to radiotherapy and the prognosis remains poor.

Discussion

Dermoid cysts are histologically benign maldevelopmental lesions which cause local symptoms as a result of their size and location.1 Rupture of the cyst wall with leakage of keratinous contents into adjacent tissues is common, usually resulting in a foreign body giant cell reaction. Malignant change of the lining epithelium into a squamous cell carcinoma is exceptional and has never previously been reported in this site. Malignant transformation appears to be a late complication of such lesions2-4 and highlights the need for the investigation and treatment of these lesions at an early stage.

The nature of the stimulus to malignant transformation in this case is uncertain; trauma is not thought to play an important role in these circumstances and there is no justification in histological terms for implicating an oncogenic virus, such as human papilloma virus. Prolonged chronic inflammation in longstanding lesions is probably important in influencing the development of a squamous cell carcinoma, as in chronic ulcers and sinus tracks in the skin. The neoplastic transformation was confined to the squamous epithelium alone in this case; the other components of the dermoid cyst showed no evidence of either dysplasia or neoplasia. The demonstrable origin of the carcinoma from the lining epithelium is against the possibility of the neoplasm representing a metastasis to this site from an occult primary and indeed there is no evidence of a primary neoplasm elsewhere in this patient.

The squamous cell carcinoma in this case was a highly invasive recurrent neoplasm in which the one year mortality can be expected to approach 100%5 The treatment of choice in such cases is
total removal with due care taken not to spill the cyst contents in the wound as this may lead to an early recurrence. Radiotherapy has not proven to be of curative value in the management of patients with advanced disease, as in this case. The outlook for this patient is poor; had the cyst been removed at an earlier stage when it was smaller in size than after the onset of malignancy, a curative surgical procedure could have been performed and the prognosis dramatically improved.

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References