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Case Report

Langerhans cell histiocytosis of the perianal region

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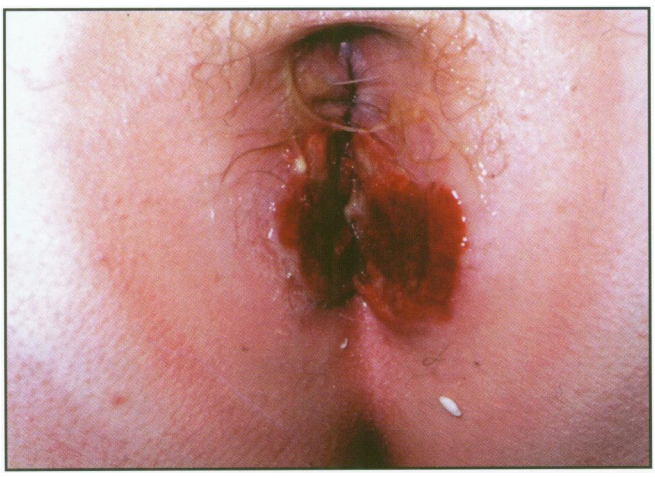
A 19 year-old male presented to the surgical outpatients with a two year history of a perianal lesion which had recently increased in size. He reported that it was only occasionally painful but his main problem was that of surface bleeding. He gave no history of trauma. However, he did report that he had had a neurosurgical procedure many years previously.

Examination showed two flat sessile lesions on each buttock in the perianal region which were granulomatous in appearance. There was evidence of surface bleeding. (figure) These findings were highly suspicious of squamous cell carcinoma of the anus. He was boarded for biopsy, which was performed without complication.

The histopathology showed ulceration of the epidermis and underlying sheets of histiocytic cells, appearances in keeping with Langerhan's cell histiocytosis or Histiocytosis X. There was no evidence of malignancy.

The lesions were subsequently excised and the wound was allowed to heal by secondary intention and did not require grafting.



Figure

When he was informed of the diagnosis, he reported that his previous neurosurgical procedure had been performed for this condition.

Examination of the archived charts confirmed this. Indeed, 11 years prior to this incident he had attended a neurosurgical unit with a two-year history of proptosis affecting the right eye. Routine neurological examination was normal except that his right eye was displaced downwards and forwards. A CT scan showed destruction of the bone in the right anterior frontal region with a soft tissue mass extending into the anterior cranial fossa, in keeping with an extra-dural mass. A right frontal craniotomy had been performed and the lesion was removed in a piecemeal fashion. It was felt at the time of the procedure that not all the lesion had been excised; however, the bony defect was repaired with titanium gauze. Two months later he underwent a course of chemotherapy, consisting of prednisolone, vincristine followed by mercaptopurine. One year later he developed diabetes insipidus. However a repeat CT scan had shown no pituitary involvement. He responded well to nasal desmopressin. A repeat CT scan six months later showed no evidence of further disease and he subsequently underwent a titanium cranioplasty to cover the bony defect. He was kept under review with no evidence of recurrence and a MRI four years later showed no evidence of recurrence of disease.

He remained well with no symptoms until his attendance at the surgical outpatients five years

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later. Since then an isotope bone scan and CT abdomen and pelvis have been normal. He remains under review.

DISCUSSION

Langerhan's cell histiocytosis (previously known as histiocytosis X) is a rare condition that generally affects children. It comprises a group of disorders that demonstrate proliferation of the Langerhan's cell derived from bone marrow.

The clinical spectrum is wide but can be generally classified according to three distinct clinical entities;

1. Sib-Letterer-Siwe disease. An acute progressive disseminated disease, which presents with fever, anaemia, organomegaly and thrombocytopenia. Death may occur due to infections or progressive anaemia.
2. Hand-Schuller-Christian disease. An intermediate clinical form *aka* multifocal eosinophilic granuloma. This often presents with the classical triad of diabetes, proptosis and bony lesions. This syndrome would indeed be attributable to the case in question.
3. Hasimoto-Pritzker disease. A congenital form of the disease that is often self-limiting.

The underlying cause is still as yet unknown. Various hypotheses have been cited as to the cause ranging from reactive processes to neoplastic processes.

The annual incidence is in the range of 5per million per year with a male: female ratio of 2:1.

The classical multifocal form, which has occurred in this patient only rarely, presents with all three of the triad of proptosis, diabetes insipidus and bony defects.

When a patient is diagnosed with Langerhan's Cell Histiocytosis, a standardised evaluation is usually undertaken; this has been developed by the Histiocyte Society. Routine blood tests should include full blood count, liver function tests and coagulation studies. Due to the possibility of diabetes insipidus a urine osmolality is mandatory, as is a chest radiograph.

More specific tests are required depending on the site of the suspected lesion.

Treatment of the condition is dependent upon the extent of the disease and upon the involved organs. Cessation of smoking is essential. Glucocorticoids have been used either topically for skin lesions or systemically for more invasive disease. Chemotherapeutic agents are indicated for multisystem disease. Trials have shown combinations of vinblastine, etoposide and prednisolone for a period of six weeks to be effective, followed by mercaptopurine, vinblastine and prednisolone for one year.

Prognosis is variable upon the type of disease encountered. Unifocal LCH generally has an excellent prognosis whereas multifocal disease has a much poorer prognosis. Letterer-Siwe disease has an even poorer prognosis and mortality can reach 50%.

Relapse is not uncommon and can occur up to 10 years after the disappearance of the original disease.

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