

Sinus histiocytosis with massive lymphadenopathy (Rosai-Dorfman disease): response to acyclovir

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We report a case of Rosai Dorfman disease which responded to acyclovir.

Case report

A 10-month-old baby girl, born in Manchester of Nigerian parents, presented with a vesicular rash and a 2 week history of submandibular and cervical gland enlargement. A varicella-zoster infection was confirmed by a rising antibody titre (CFT) of greater than 1/80. Following resolution of the rash, cervical and submandibular swelling increased with mild axillary and inguinal involvement, splenomegaly and a low grade pyrexia. Chest radiographs revealed right middle lobe infection with paratracheal nodal enlargement. Penicillin and flucloxacillin were ineffective.

Initial investigations were unhelpful: lymph node biopsy showing reactive changes only.

By 13 months the massive lymphadenopathy was causing neck retraction, airway compromise, irritability and failure to thrive.

Reinvestigation revealed a haemoglobin of 8.0 g/dl. The peripheral white cell count was $21.2 \times 10^9/l$, absolute neutrophil count $16.1 \times 10^9/l$ and the platelet count was $976 \times 10^9/l$. Absolute T4 count, T4 helper, CD4 and C reactive protein were normal. The erythrocyte sedimentation rate was 41 mm in the first hour. There was a polyclonal hypergammaglobulinaemia with IgG 21 g/l (normal range 2.5-11.0 g/l), IgM 3.2 g/l (0.3-2.0 g/l) and IgA 0.5 g/l (0.2-1.0 g/l). Paul Bunnell result was positive but Epstein-Barr capsid antigen negative. HIV antibody was negative. Urea and electrolytes, liver function tests and a coagulation screen were all normal as were skeletal survey, bone scan and abdominal ultrasound, skin biopsy and bone marrow. Differential Mantoux and prolonged culture of first lymph node were both negative. A repeat lymph node biopsy now revealed features of Rosai-Dorfman disease.

The worsening clinical condition necessitated treatment, and in view of the initial varicella-zoster infection, a trial of oral acyclovir, 100 mg five times daily, was commenced. Within 4 weeks there was a marked reduction in lymph node size and the total white cell count was $11.6 \times 10^9/l$. Treatment was stopped after 11 months as she was thriving (Figure 1). There has been no recurrence of the condition clinically after 3 years follow up.

Discussion

Rosai-Dorfman disease (SHML) was established as a diagnosis following two reports in 1969 and 1972^{1,2} and 423 patients in the SHML registry have recently been reviewed³.

SHML is a disease of uncertain aetiology, classified by consensus, as an idiopathic histiocytosis with a varied clinical presentation, and specific histological features. The usual presentation is of massive cervical lymphadenopathy with fever, weight loss, malaise or night sweats occasionally

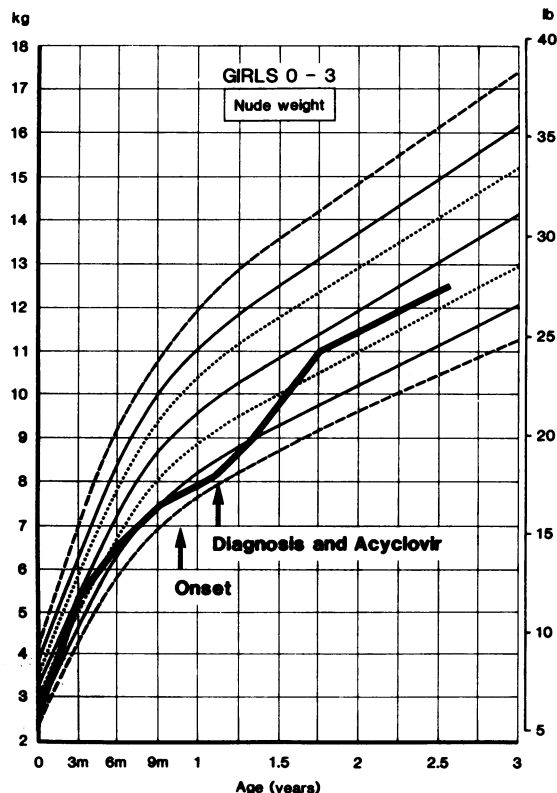


Figure 1. Growth chart showing catch up growth after treatment with acyclovir

noted. There is often a raised ESR, anaemia and neutrophilia with polyclonal hypergammaglobulinaemia. Nearly one half have extranodal disease.

Nonspecific immune involvement was found in 56 of the 423 registry patients and 11 cases presented with malignancies. Positive virological serology was found in 10% the most common agent being Epstein-Barr virus.

In the 238 registry patients who had a one year follow up spontaneous resolution occurred in 20%. One half had persistent but stable disease, 15% possibly had persistent disease and three patients had progressive disease. Twenty-one patients had died, four of Rosai-Dorfman, 13 with evidence of the disease persisting and four free of the disease.

There are reports of treatment having been used in 84 patients. Thirty-nine patients had surgery for life or function threatening disease. Ninety-four courses of chemotherapy were given. The best response rates were for a combination of vinca alkaloid plus alkylating agent and corticosteroid with 7 out of 13 (53%) responding. Radiation therapy gave a complete or partial response in only 10 of 34 (29.4%) patients treated.

Acycloguanosine (acyclovir) has never been used before in this condition. The rapid improvement in our patient was thought to be a response to treatment rather than spontaneous resolution. There are theoretical reasons why acyclovir may work in other cases of Rosai-Dorfman disease. It is effective against Epstein-Barr virus⁴⁻⁶ and may have cytotoxic effects on host cells as an inhibitor of DNA polymerase. We suggest that acyclovir be considered as an early treatment in future cases in order to assess its efficacy further.

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Obturator hernia: an elusive diagnosis

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Strangulated obturator hernia is an uncommon entity, but is potentially lethal for the elderly population it usually affects. The clinical picture is often confusing. The following is a case report of a very atypical presentation of an obturator hernia.

Case history

An 80-year-old, thin, frail woman was admitted with a large abscess on the medial side of her left thigh. She came from a nursing home where she was confined to bed and suffered from senile dementia. Approximately 2 weeks prior to admission she had an episode of vomiting which settled spontaneously. For 5 days before admission she complained of increasing pain in her left thigh on movement. Her oral intake dropped, though she was moving her bowels. She was dehydrated and toxic with a temperature of 39.6°C.

On examination, she had a large abscess in the medial side of her left thigh with redness and induration extending over a wider area. There were tender, mobile lymph nodes in the left groin and her abdomen was soft with normal bowel sounds. Her leucocyte count was $39.5 \times 10^9/l$, blood urea nitrogen was 28 mmol/l and plain abdominal X-rays suggested localized ileus over the left iliac fossa.

Incision and drainage of the abscess was performed under sedation, evacuating 100 ml of dark, foul smelling fluid. She was commenced on antibiotics.

Her condition improved rapidly, her temperature and blood indices returned to normal and her appetite and mental state improved also. Her wound continued to drain, necessitating two to three dressing changes per day for the first 2 weeks. Per- and postoperative swabs of the draining fluid cultured mixed intestinal flora. A barium meal follow through performed one week after admission showed a knuckle of small bowel caught at the left obturator foramen with a fistulous tract into the canal, without obstruction (Figure 1). The discharge from the thigh wound gradually dried up and the patient was discharged one month after admission.

Discussion

Obturator hernia is usually unsuspected and hence undiagnosed before an exploratory operation. There are four cardinal features of the condition which may or may not be present together. They are: (1) the Howship-Romberg sign, (2) intestinal obstruction, (3) previous attacks of bowel

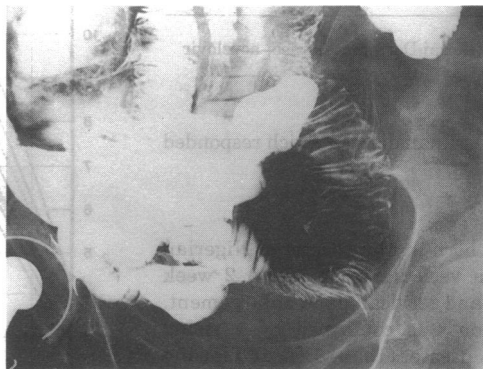


Figure 1. Barium meal showing a knuckle of small bowel caught at the left obturator foramen with a fistulous tract into the canal

obstruction resolving spontaneously, (4) a palpable mass in the groin with the patient supine, thigh flexed, adducted and rotated laterally. Other rare signs reported in the literature are: (a) ecchymosis on the medial part of the groin below the inguinal ligament in a patient who does not have a femoral hernia¹, (b) the finding of a tender mass over the obturator area on vaginal or rectal examination¹ and (c) subcutaneous emphysema of the thigh².

On reviewing the literature we found only four cases where strangulated herniae presented as an abscess in the thigh³⁻⁶. This is only the third one in which an abscess was the only presenting feature with no signs of intestinal obstruction^{4,5}. The history suggests that she had a Richter's type strangulation about 2 weeks before, which later perforated into the thigh leading to abscess formation. The resulting fistula closed spontaneously in the following weeks, leading to a full recovery, which was fortunate for our patient in view of her physical condition.

The presence of obstruction with a thigh abscess should suggest strangulation in the obturator canal if femoral and inguinal hernia can be ruled out. It is a very difficult condition to diagnose and even more so when a patient cannot give a good history due to senility or confusion as in this case.

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