Case reports

Juvenile pancreatic pseudocyst

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Pancreatic pseudocysts in children are rare, and usually require surgical treatment. We report a post-traumatic juvenile pancreatic pseudocyst that has been successfully treated without operation.

Case report

An 8-year-old boy sustained blunt trauma to the abdomen after being crushed between a car bumper and a garage door. On admission to hospital he was tender across the epigastrium but was otherwise well, although he had recently contracted mumps with characteristic bilateral parotid swellings.

The serum amylase on admission was 387 iu/l, and an abdominal ultrasound examination demonstrated free fluid around the inferior aspect of the liver. A CT scan, however, showed no intra-abdominal abnormality. The child's condition settled over a 3-day period, and his serum amylase returned to normal. He was discharged from hospital, but one month later presented with a 48-hour history of nausea and epigastric pain. On examination he was tender over the left upper quadrant of his abdomen where a firm mass was palpable which did not move on respiration. His serum amylase was 787 iu/l, and both an ultrasound examination and a CT scan demonstrated a 10×7 cm cystic lesion arising from the tail of the pancreas (Figure 1).

Over the subsequent 3 days the child was asymptomatic, and a decision for conservative treatment was therefore taken. Ultrasound examination at monthly intervals thereafter demonstrated progressive decrease in size of the pancreatic pseudocyst, and 5 months after the original injury the child remains fit and well with no clinical or ultrasonic evidence of an intra-abdominal mass.

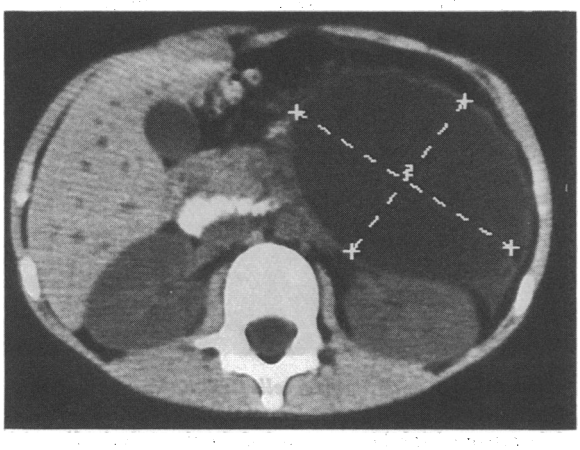

Figure 1. CT scan demonstrating a 10×7 cm cystic mass arising from the tail of the pancreas

Discussion

Prolonged observation of adult patients with pancreatic pseudocysts in anticipation of spontaneous regression carries a high risk of complications when compared with surgical management[1,2]. Operative treatment is also preferred for children with pancreatic pseudocysts, and a conservative policy is reserved only for those in whom associated symptoms improve, with a return to normal serum amylase levels, and resolution of the pseudocyst as determined by sonography[3].

Pseudocysts of the pancreas are uncommon in persons under the age of 18 years; just 113 cases were documented in a 1981 review of the literature with a mean age for presentation of 7.5 years[3]. Boys are more commonly affected than girls, perhaps because they are more prone to trauma: more than half of all cases described have a history of trauma, usually caused by handlebar injuries or by child abuse[4]. The pancreas of the child described here may have been affected by the mumps virus prior to the episode of blunt abdominal trauma and this may have exacerbated the subsequent pancreatic inflammation[5].

Common presenting signs and symptoms of pancreatic pseudocysts in children include abdominal pain, nausea, vomiting, anorexia and weight loss, and an abdominal mass has been found in up to 64% of patients[6]. Up to 80% of patients have a raised serum amylase at presentation[6]. Ultrasonography and, more recently, CT scanning are the preferred methods of investigation, but ultrasound examination may be limited after blunt abdominal trauma by the presence of gas[3,7].

Although occasional success with nonoperative treatment of juvenile pancreatic pseudocysts has been reported[4], surgical intervention is considered the best mode of therapy. Procedures used have varied from total excision of the cyst, to external drainage or marsupialization, but the most common operation used has been internal drainage through a cystogastrostomy, which, as in adult patients, has the lowest associated recurrence and mortality rates[3,4]. Although percutaneous cystogastrostomy guided by ultrasonography and gastroscopy has recently been used in adults[8], the use of this technique in children has not been described.

Juvenile pancreatic pseudocysts have an excellent outlook for cure. Although there should be no hesitation in operating on children with persistent abdominal pain, vomiting and fever or on those with an elevated serum amylase with failure of resolution of the cyst, it appears that a conservative policy can be safely adopted if such features are absent, even when a cyst is very large, as in the case described.

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Primary small bowel ganglioneuroblastoma and Friedreich's ataxia

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A primary ganglioneuroblastoma of the small bowel is an extremely rare occurrence. We describe a patient with the hereditary spinocerebellar degeneration of Friedreich's ataxia, who was found to have a primary small bowel ganglioneuroblastoma during pregnancy. The literature is reviewed with particular regard to extra-adrenal neuroblastoma and any relationship to congenital disease.

Case report

A 26-year-old woman attended the antenatal clinic 30 weeks pregnant. She was mentally subnormal and Friedreich's ataxia had been diagnosed in late childhood. She was ataxic and dysarthric with a marked kyphoscoliosis, pes cavus and optic atrophy of the right eye. Her mother also suffered from Friedreich's ataxia.

At this first visit she was hypertensive and was admitted for bed rest and control of her blood pressure. This, however, proved unresponsive to treatment and it was necessary to perform a caesarean section at 35 weeks maturity. A healthy female child was delivered. During closure of the abdominal wall a loop of bowel presented at the wound with a solid oval tumour on the serosal surface. The lesion, 2 × 1 cm, was removed by shaving it from the serosa. She made a good recovery and the blood pressure returned to normal. Histology of the tumour showed it to be a malignant ganglioneuroblastoma.

The hydroxymandelic acid level postoperatively was 17.9 μmol/24 hours, the normal for this laboratory being 10–35 μmol/24 hours. Radiological investigation showed no evidence of distant spread. There was no history of diarrhoea or flushing such as may occur in excessive catecholamine production.

A laparotomy six weeks after the caesarian section showed there was a small area of residual tumour on the serosal surface of the antemesenteric border of the mid small bowel, with metastases in the lymph nodes in the adjacent mesentery. A detailed but negative search was made of the more usual sites for neuroblastoma. The tumour and mesentery were resected and she made an uneventful recovery. She remains well with no evidence of residual or metastatic disease 18 months after operation.

Pathology: The specimen removed at caesarean section measured 2.5 × 2.5 × 2.0 cm and had a grey external surface studded with small pale nodules up to 0.2 cm in diameter. On the cut surface the lesion appeared to be encapsulated except for one area which was presumed to be the site of attachment to the small intestine. The cut surface showed areas of dark, crumbly, partly necrotic, haemorrhagic tissue and areas of firmer grey-white tissue.

Microscopically the lesion had the structure of a ganglioneuroblastoma (Figure 1). The haemorrhagic areas consisted of sheets of fairly uniform, rounded or oval nuclei with scanty, poorly defined, eosinophilic cytoplasm. Mitoses were common. In the firmer areas the tissue was broken up by a network of capillaries. In these areas there was a background resembling glial fibres and some of the cells were arranged in rosettes or pseudo-rosettes. Larger cells resembling ganglion cells were present in these areas. The nodules on the external surface of the lesion consisted of florid decidual reaction.

Some of the tissue was reprocessed for electron microscopy. No dense core secretory vesicles were identified, possibly because of the inappropriate initial fixation in buffered formal saline, but neural elements were identified in the ganglioneuromatous areas.

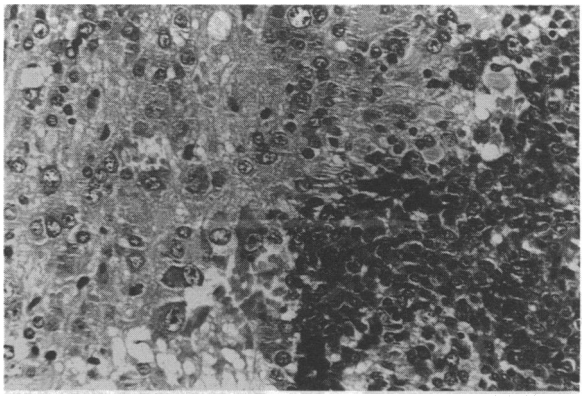

Figure 1. Large ganglion-like cells (left) have abundant cytoplasm and prominent nuclei. The neuroblastomatous element of the tumour (right) is composed of closely packed cells, with small round or oval nuclei and poorly defined cytoplasm. (H&E × 925; reduced 58%)

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