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Case report

An unusual case of abdominal pain in a patient with cystic fibrosis

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Abstract

We report the case of a 19 year old man with cystic fibrosis (CF) who presented with atypical abdominal pain precipitated by pressing his ribs. This was subsequently discovered to be referred pain from an intercostal schwannoma. Surgical resection led to a resolution of his symptoms.

Keywords: Cystic fibrosis; Abdominal pain; Referred pain; Schwannoma

1. Case report

An otherwise well 19 year old man with cystic fibrosis was seen in clinic complaining of right sided upper quadrant abdominal pain. The pain was unrelated to food, nor associated with nausea or vomiting. He denied any change in bowel habit or any history of jaundice. He regularly took pancreatic supplements and denied steatorrhoea. His lung function was well preserved with a forced expiratory volume in 1 s (FEV_1) of 4550 ml and forced vital capacity (FVC) of 5500 ml. Examination revealed a soft non-tender abdomen, without organomegaly or other masses. However, on repeat questioning, the patient confessed to getting the pain on being startled by someone touching him unexpectedly. On further examination, the pain could be reproduced by firm pressure over the right chest wall. Blood results including full blood count, urea and electrolytes, bone profile and liver biochemistry were unremarkable. Erythrocyte sedimentation rate (ESR) and C reactive protein (CRP) were also normal.

A chest radiograph showed bronchiectasis consistent with his known cystic fibrosis. There was no evidence of rib fracture. An abdominal radiograph was unremarkable. A bone scan was normal but a chest wall ultrasound revealed a $13 \times 30 \times 15$ mm well-defined intercostal neuroma.

The tumour was localised by ultrasound and removed under local anaesthesia. The histology (Fig. 1) confirmed a well-circumscribed 3.1 cm × 1.4 cm neurilemmoma (schwannoma) with some degenerative changes. The lesion was benign and completely excised with an intact capsule. The patient is now asymptomatic.

2. Discussion

With improving life expectancy gastrointestinal complications are becoming an increasingly important cause of morbidity in patients with cystic fibrosis [1]. The causes of right upper quadrant pain are similar to the general population. However, a number of gastrointestinal complications are more commonly seen in CF patients, related to the generalised increased viscosity of their secretions. Distal intestinal obstruction syndrome is the retention of viscous material in the colon leading to varying degrees of bowel obstruction and occurs in up to 16% of patients [2]. There is also an increased incidence of gallstone formation and cholecystitis in CF patients due to the production of more concentrated bile. Reflux oesophagitis, pancreatitis in those with residual function, fibrosing colonopathy associated with certain pancreatic enzyme preparations and renal colic from

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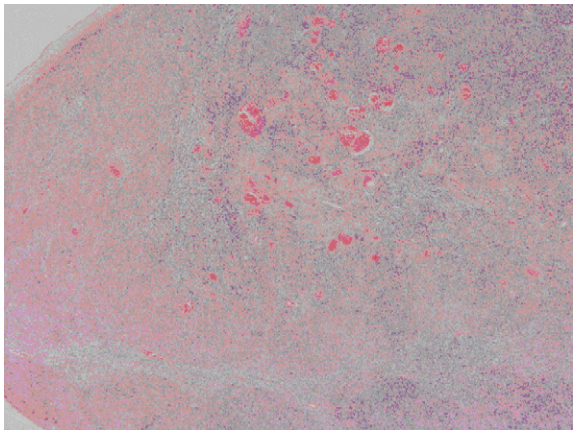

Fig. 1. Haematoxylin and Eosin, ×20: circumscribed encapsulated tumour with randomly admixed hyper and hypocellular areas and ectatic blood vessels typical of schwannoma.

stone formation are all recognised causes [1]. It would have been tempting to consider any of these as the likely cause of this patient's pain. However, the pain was not characteristic of any of these conditions. It seemed more consistent with impingement of an intercostal nerve leading to referred abdominal discomfort.

Myelin is produced by schwann cells and provides covering for nerve roots. Schwannomas are usually benign tumours arising from these cells [3]. They are generally slow growing and can cause symptoms by stretching the nerve. Their biological behaviour is similar to neurofibrommas except invasion of the nerve is uncommon. The commonest form is an acoustic neuroma arising on the vestibular division of the VIIIth cranial nerve. Intercostal neurogenic tumours are relatively uncommon. Less than 10% of primary neural tumours of the chest originate peripherally from intercostals nerves, the majority being found in the mediastinum [4]. They are usually asymptomatic and are found incidentally on chest radiographs [5]. Treatment is curative, as in this case, with surgical resection.

We suggest that in CF patients with an atypical presentation of abdominal pain, referred causes should be considered.

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