Idiopathic thrombocytopenic purpura: a case of particular historic interest

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Idiopathic thrombocytopenic purpura (ITP) is a disease in which the blood platelets are affected by autoantibodies and are removed from the bloodstream by the spleen and other reticuloendothelial tissue. The resulting bleeding disorder may be a transient sequel to childhood viral infections, but in other circumstances it is a prolonged and serious disease of children and adults. A case is reported of the first successful treatment of chronic ITP in England. It highlights the characteristics of the disease and an unusual complication.

Case report
At the age of 5 years in 1921 the patient was well except for easy bruising. She suddenly presented to medical attention with severe haematemesis, melaena and epistaxis. A widespread impalpable purpuric rash was present, and the spleen was enlarged. The bleeding time was prolonged to ten minutes though the clotting time was normal, and the blood film showed a very severe thrombocytopenia. During the next three years a further eight episodes of severe bleeding occurred, and between these episodes purpura, bruising and epistaxis were the rule. Finally the patient became bedridden with persistent nasal oozing, anaemia, mild jaundice and shortness of breath on exertion. The platelet count at this time was 5 x 10⁹/l and the haemoglobin 40% of normal.

The child's attendant physicians, Sutherland and Williamson, knew that in 1915 Kaznelson had removed the enlarged spleen of a patient with ITP and that remission had followed (Kaznelson 1916). They decided that splenectomy might offer a chance of improvement to their patient. During a period when there was no active bleeding a spleen of two-and-a-half times normal size was removed uneventfully and in the following weeks the platelet count rose to 35 x 10⁹/l (Sutherland & Williamson 1925). In spite of episodes of purpura and severe menorrhagia at the menarche, the patient enjoyed a partial remission and led a normal life with no further major bleeding episodes.

In adulthood the platelet count was usually 12–20 x 10⁹/l and sparse purpuric lesions persisted. A hiatal hernia was diagnosed by X-ray and mild hypertension was controlled by hydrochlorothiazide (Moduretic). Two gynaecological operations resulted in severe bleeding, but an extraction of a tooth was uneventful. Howell-Jolly bodies were noted in the blood film but were absent from 1972 onwards.

In 1979 the patient noticed extensive purpura and easy bruising. She had a small epistaxis. The platelet count was 5 x 10⁹/l and the blood film was normal. Because hypertension and the hiatal hernia increased the risk of bleeding, corticosteroids were administered. On prednisolone 40 mg daily the symptoms improved and the platelet count rose to 18 x 10⁹/l but her blood pressure increased and she began to notice dyspepsia. On reducing the dose of prednisolone severe purpura recurred.

Because the blood film showed no changes of splenectomy an isotope spleen scan was carried out and this showed a splenunculus in the splenic bed. Splenunculectomy was carried out and 10 units of platelets transfused during the operation. The splenunculus weighed 29 g and had normal histology. Two weeks after the operation the platelet count was 17 x 10⁹/l, and it remained elevated after prednisolone was discontinued. Howell-Jolly bodies reappeared in the blood film. However, a few weeks after the prednisolone was completely discontinued, bruises had reappeared and she was suffering from steroid withdrawal symptoms. Prednisolone had to be re-started and the patient is now well, with bruising controlled, on a daily dose of 10 mg. The last platelet count was 25 x 10⁹/l.

Discussion
Since Kaznelson demonstrated the effect of splenectomy and postulated that the spleen sequestered and destroyed platelets, the only major advance in the understanding of ITP has been the demonstration of platelet antibodies (Veenhoven et al. 1980). These are demonstrable both attached to platelets and free in the serum of sufferers, though all attempts to define a specific antigen have failed. It has been impossible to predict the response to splenectomy, either by measuring the uptake of radio-labelled platelets by the spleen, or by noting the type of response to corticosteroid therapy (Richards & Thompson

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Complications of a large pharyngeal pouch

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This case is reported because of an unusual complication, which occurred after the surgical treatment of a large pharyngeal pouch.

Case report
The patient was a 57-year-old agricultural worker. He presented with a five-month history of regurgitation, dysphagia and weight loss. Endoscopy and contrast studies had been performed at another hospital three weeks earlier and six days before admission he had developed fever, shortness of breath and a nonproductive cough.

Examination revealed a sick, emaciated man with a large left-sided pleural effusion. A gastrograin swallow demonstrated a massive pharyngeal pouch, with no spill into the pleural cavities or visualization of the distal oesophagus (Figure 1). The empyema was drained and sterilized by repeated instillation of benzylpenicillin, cultures having grown Streptococcus milleri.

Three weeks later the neck was explored through a collar incision and the pouch was packed with gauze, mobilized and amputated with a myotomy of cricopharynx. The pharyngeal defect was closed with a double layer of catgut. The empty pouch measured 12 × 8 cm.

Postoperatively a soft cystic swelling developed deep to the cervical incision and enlarged to 12 cm in diameter. It was air-filled and inflated when the patient blew against pursed lips, but not when a Valsalva manoeuvre was performed. On the eighth postoperative day a second gastrograin swallow revealed an air-filled cavity or aerocele in the superior mediastinum, and in communication with the pharynx (Figure 2). The patient remained well without any signs of mediastinitis but was not permitted to eat or drink. A third gastrograin swallow on the sixteenth postoperative day did not show any entry of contrast into the aerocele and the patient began to drink without adverse effects.

The aerocele remained unchanged in size or position for several weeks, with the patient eating normally until an episode of fever, local inflammation and purulent oze from the cervical incision was followed by its complete disappearance.

Discussion
The case is instructive for several reasons. Firstly, the pharyngeal pouch was associated with the development of a large empyema which, despite its size, had produced symptoms for only a few days, suggesting that it had arisen rapidly either

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