Medical Memoranda

Argentaffin Tumour of the Ileum with Intussusception

According to Dangremond (1942), only 283 argentaffin tumours of the small intestine had been recorded in the literature since they were first recognized as a distinct entity by Oberndorfer (1907).

Ariel (1939), in a review of the cases published between 1930 and 1938, was able to record the age of the patients in 70 instances, and their disposition in decades was as follows :

0-10 Nil	31-40 6	61-70 17
11-20 Nil	41-50 9	71-80 9
21-30 1	51-60 27	81-90 1

He points out that the average age of 57.5 years differs greatly from that of the patients admitted to the Mount Sinai Hospital with appendicular tumours, whose average age was only 25. He attributes this to the much earlier onset of symptoms in the appendicular cases. This contrast had also been made by Humphreys (1934). Macfarlane (1957) has stated that in his own series of non-appendicular cases the extremes of age were 19 and 83.

The occurrence of one of these tumours in the ileum in a boy of 4 years is therefore of considerable interest. It has long been known that these tumours grow extremely slowly, and according to Cope (1930) may attain a large size and then remain stationary for years. Many cases may therefore originate in infancy or childhood, and we believe our case to be the first to confirm what has previously been a speculation.

CASE HISTORY

A boy of 4 years was admitted to hospital on September 22, 1957, with a three-days history of generalized abdominal pain, diarrhoea, and repeated vomiting. He was flushed, his tongue was coated, and his abdomen was soft and distended. Rectal examination showed the presence of watery altered blood, and x-ray examination showed gaseous distension of the small gut and a fluid level in the stomach.

Operation.—At laparotomy an ileo-colic intussusception was found. This was reducible except for the last 4 in. (10 cm.) of the ileo-ileal segment. This portion was resected and an end-to-end anastomosis carried out. Recovery was uneventful.

Pathology.—The resected intussusception was firmly fixed and irreducible. At its apex there was a firm white submucous nodule 1 cm. in diameter. The gut was purple from the extravasated blood and the mucosa was coated by fibrinous exudate. Histological sections showed the tumour lying between the muscle coat and the degenerating mucosa. It was composed of islands of solidly packed anaplastic cells with no structural arrangement and separated by a delicate fibrous stroma. There was no capsule. In sections stained by Levaditi's method the cells were seen to be packed by grey-brown silver-retaining granules.

COMMENT

These tumours come to light in six different ways: (1) As unexpected findings at necropsy. Dangremond's series of 46 included 23 found this way. All the six

recorded by Willis (1940) were necropsy cases, as were the nine described by Raiford (1933). At least half the published cases appear to have been "pathologist cases" and had presumably been symptomless. (2) In the operative treatment of acute or subacute obstruction. No fewer than 33 of Ariel's 79 review cases fall into this category. This is the usual clinical manifestation. (3) A large tumour may undergo a central necrosis and lead to perforation. This is rare, but cases have been described by Cooke (1931) and Stewart and Thomson (1951). (4) A nodular growth may lead to intussusception as in our case. In Ariel's review series it was seen once in a man of 52, and among the 11 cases in Ariel's personal series there was one instance in a man of 32. This complication is unlikely once the tumour has reached a certain size because of its diffusive infiltration of the mucosa and the muscle. (5) The discovery of a painless mass in the abdomen during life. (6) If the tumour, together with its metastases, is bulky enough the secretion of 5-hydroxytryptamine may lead to the clinical syndrome of flushing, with cyanosis, diarrhoea, and other symptoms described by Björck and his colleagues (1952).

Consideration of these six groups makes it evident that in a child of 4 years, apart from the improbable event of a small nodule being found at necropsy, by far the most likely way for the tumour to come to light would be in relation to an intussusception. From this it follows that the apex of any intussusception in a child should be rigorously searched. In our case, had the surgeon not drawn attention to what appeared to be a scar in the region of the tumour the pathologist would almost certainly have overlooked it. The swollen, distorted, and discoloured gut will of course obscure small submucosal tumours.

The presence of other neoplastic centres or of metastases cannot be ruled out. According to Macfarlane the value of estimating 5-hydroxyindole acetic acid (the end-product of 5-hydroxytryptamine) in the urine, after surgical removal of a tumour, in terms of its ability to indicate early metastases, has not yet been established. The urine in our case was examined a month after the operation and no significant rise found. Logically the patient should be brought to hospital for the urine to be examined every year or so, but the psychological damage which might result in the future should be carefully weighed before a lifelong observation is undertaken.

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