Ectopic thymus of the neck—report of three examples in children

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Summary

Three children with ectopic thymic tumours of the neck are described. One solid mass was situated in the midline whilst two were unilateral and cystic. The difficulties of diagnosis and surgical removal are stressed and the question of interference to immunological competence discussed.

SMALL lobules of aberrant thymus are not infrequently found in the neck, often within the thyroid gland, possibly present in up to 20% of humans (Goldstein and Mackay, 1969; Gilmour, 1941). However, ectopic thymic tissue of size sufficient to produce a palpable swelling of the neck is uncommon. In a review of the forty-six such cases recorded in the English literature, Hinds *et al.* (1970) found that the majority were diagnosed in childhood.

Case 1

A 7-year-old girl was noticed to have a tumour of the neck from the age of 6 months. It increased in size gradually but produced no symptoms. On examination, there was a midline, apparently fluctuant lump which moved on swallowing but had no superficial fixation. The pre-operative diagnosis lay between a midline epidermoid cyst and a thyroglossal cyst.

At operation a solid mass with no obvious vascular supply lay deep to the infrahyoid muscles and extended inferiorly behind the sternum, its superior pole being the palpable midline swelling (Fig. 1). It proved to be a discrete lobulated structure about 5 cm long and 1.5 cm wide which was shown microscopically to be composed of normal thymic tissue.

A lateral tomogram 4 weeks after surgery demonstrated an apparently normal retrosternal thymus.

Case 2

An 8-year-old boy presented with a swelling of the left side of the neck which had been present for one month. On examination, this clinically euthyroid child had a firm ovoid mass which moved on swallowing and appeared to be an integral part of, if not the entire, left lobe of thyroid. Thyroiditis and carcinoma of the thyroid were considered as possible diagnoses.

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FIG. 1. Case 1. Discrete mid-line thymic tumour being delivered through an incision about 50 mm above the sternal notch.

At operation, a thin-walled cyst, quite separate from the thyroid, was adherent to the infrahyoid muscle and the carotid sheath from which it was removed completely with considerable difficulty. It weighed 29 g, measured 5.8 × 2.8 cm and contained yellow fluid with visible cholesterol crystals. Its wall was 0.3–0.5 cm thick and included several smaller cysts. Microscopically each cyst was lined with a single layer of flat endothelium: the wall was fibrous and included areas of involuted thymus and collections of cholesterol crystals some of which were associated with a foreign body giant cell reaction.

Case 3

A 12-year-old boy presented with a 3-month history of a swelling in the right side of his neck which initially had been painful and recently had increased in size.

On examination an indiscrete cystic mass measuring about 3 × 2 cm was palpable anterior to the sternomastoid muscle. It was diagnosed with confidence as a branchial cyst.

At operation it extended from the angle of the mandible, deep to the sternomastoid muscle and close to the internal jugular vein whilst the vagus nerve coursed through it. It consisted of a number of cysts containing gelatinous yellow fluid and cholesterol crystals. The cysts were lined with cells

FIG. 2. Case 3. Wall of cyst showing cellular lining and surrounding thymic tissue.

resembling those found normally in Hassell's corpuscles and the solid elements of the mass consisted of degenerate thymic tissue (Fig. 2). A normal parathyroid gland was included.

Discussion

The thymus gland is derived from the third and fourth pharyngeal pouches as paired structures in the fourth week of intra-uterine life which migrate caudally to a position between a sternum and heart and separate from the pharynx whilst the cephalic rudiments disappear (Warkany, 1971; Arey, 1965). Speer (1938) suggested that retention of thymic tissue in the neck could be explained in various ways. These included: (a) proliferation of isolated remnants of branchial cleft, thymo-pharyngeal ducts or thymic tubules; (b) failure of the cephalic processes of the elongating thymus to involute; (c) neoplastic proliferation of cephalic remnants (Wadas, 1934); (d) overgrowth of the connective tissue stroma involved in the development of the thymus. Hinds *et al.* (1970) stated that 95% of thymic tumours in the neck were unilateral and that 90% were cystic. The difficulty of pre-operative diagnosis is illustrated by the variety of suggested diagnoses in the three patients presented. These examples, particularly cases 2 and 3, show that the mass may extend widely and deeply in the tissues of the neck and may involve important structures, rendering surgical removal considerably more difficult than is generally reported in the literature.

Interference with the immunological status quo is obviously an important consideration when thymus tissue is removed from a child. In fact, only one case had lateral tomography specifically undertaken and this suggested the remaining presence of normal thymus, as would probably be the usual case. Even if, however, the removed portion was the entire functioning thymus, it is unlikely that this would interfere with immunological competence as man appears to be immunologically more mature at birth than the experimental animals normally used for investigation (Goldstein and Mackay, 1969) and, indeed, total thymectomy has been undertaken in neonates and young children without apparent interference with the mechanism (Gotoff, 1968; Zollinger *et al.*, 1964) although the theoretical risk remains of subsequent predisposition to neoplasia in the long term (Waksman, 1964; Goldstein and Mackay, 1969).

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Renal failure and suprarenal calcification after secondary haemorrhagic disease in a newborn baby

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Summary

A newborn baby who was severely asphyxiated at birth developed secondary haemorrhagic disease which was treated successfully with intravenous heparin. However, she was later found to have supra-renal calcification, chronic renal insufficiency and hypertension. It is felt that prompt treatment of conditions that predispose to secondary haemorrhagic disease is very important as, once this has occurred, treatment may not prevent permanent damage to vital organs.

Introduction

Secondary haemorrhagic disease of the newborn occurs mainly in association with severe birth asphyxia, hypothermia and rhesus haemolytic disease (Chessells and Wigglesworth, 1970) and often results from disseminated intravascular coagulation (Boyd, 1967). Treatment with heparin (Whaun, Ormson and Oski, 1971) or exchange transfusion (Gross and Melhorn, 1971) has been advocated, but the mortality is 60% (Whaun *et al.*, 1971). Although fibrin deposits have been found in many organs at autopsy (Boyd, 1967) we have not found reports of permanent damage to organs other than the brain in the survivors (Bryant *et al.*, 1970).

Case report

This baby girl was born at 42 weeks' gestation after an antepartum haemorrhage by normal vertex delivery in a nearby maternity home and weighed 3.85 kg. She failed to breathe and after 10 min was treated with positive pressure ventilation via an endotracheal tube, regular respiration being estab-

lished at 40 min. She was then transferred to the Special Care Baby unit. On arrival she was moribund, with no spontaneous movements, apart from shallow respiration, no primitive reflexes and heart rate of 90/min. Sodium bicarbonate was given intravenously and she was nursed in 59% oxygen. Her heart rate rose to 140/min, respiration improved and some spontaneous movements occurred. Investigations gave the following results: Hb 11.7 g/100 ml, platelets 103,000/mm³, prothrombin activity 12%, pH 7.29, Pco₂ 38 mmHg, Po₂ 180 mmHg, and serum bicarbonate 18 mEq/l. She was given 80 ml of blood and 1 mg of vitamin K₁.

The following day her haemoglobin had fallen to 10.6 g/100 ml and she was given a further blood transfusion. Over the next 4 days she had a number of fits which were treated with phenobarbitone. Her general condition improved with increase in activity and the Moro reflex returned, although she could not suck and had marked head retraction and severe hypotonia. Her urine contained a large amount of blood, but was not infected and the kidneys were not enlarged. On the sixth day of life her haemoglobin had fallen to 8.4 g/100 ml, platelets to 12,000/mm³, and she bled profusely from a heel prick. A further blood transfusion was given and later that day exchange transfusion with fresh, heparinized blood was carried out. After this her haemoglobin concentration rose to 13.8 g/100 ml and platelets to 115,000/mm³.

However, the next day the haemoglobin and platelet count fell again and fibrin degradation products were detected in the serum. After another