

Ectopic thymus in the neck; a case report and review of the literature

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Summary—A soft, poorly defined mass in the right upper neck of a 7-week-old boy was shown on histology to be ectopic thymus. As aberrant thymic tissue often does change into cysts or neoplasms removal is the treatment of choice. Its persistence in the upper neck seems to be very rare. Embryology, incidence, differential diagnosis and treatment are discussed with a review of the literature.

Human embryology with its rare developmental disturbances can be a source of both puzzle and surprise for a surgeon.

no parathyroid glands on serial sections. Healing was uneventful and, with the contralateral thymus in normal position, T-cell counts and immunological functions were normal.

Case report

A 7-week-old white male presented with a swelling of his right neck. The mother had first observed this tumour soon after birth and reported that it had grown larger recently.

The healthy looking boy showed an asymmetry of his neck (Fig. 1) with a very soft, non-tender mass approximately 5 cm in diameter below and behind the right angle of the mandible (Fig. 2). It was somewhat difficult to delineate exactly the outline of the tumour; there was not the slightest hint of fluctuation and the overlying skin showed no discolouration.

A cystic tumour like a branchiogenic cyst seemed very unlikely in view of the difficulty in defining the border and the absence of fluctuation. A vascular tumour was the most likely diagnosis, probably a cervical hygroma or a lymphangioma, with a haemangioma being less likely because of the lack of discolouration of the skin. Surgical exploration was considered necessary and was performed under general anaesthesia.

The solid, lobulated tumour deep to the platysma muscle was easily dissected and seemed to consist of lymphatic tissue. At this stage a malignant lymphoma was considered a possible diagnosis, therefore no frozen section was performed and the mass was removed completely (Fig. 3).

The hypoglossal nerve ran through the tumour and was dissected free. The cavity at the end of the operation revealed the sternocleidomastoid muscle with the greater auricular nerve, the digastric muscle and submandibular gland, the hypoglossal nerve, the bifurcation of the carotid and the deep jugular vein.

Histology showed normal thymic tissue (Fig. 4) with



Fig. 1

Figure 1—Soft swelling of right upper neck in a 7-week-old male.



Fig. 2

Figure 2—Dotted line shows extensions of tumour in relation to lower border and angle of mandible immediately after removal of cervical mass.



Fig. 3

Figure 3—The 4.5 x 5 x 1.5 cm lobulated soft tumour.

Discussion

The thymus derives from the anterior extensions of the third, and possibly the fourth, pharyngeal pouch in the lateral pharyngeal wall just above the larynx. It loses its connection with the parapharyngeal wall in the sixth week and by the eighth to ninth week has descended into the mediastinum. The parathyroid glands which develop from the posterior extensions of the same pharyngeal pouches accompany the thymus on part of the descent (Warkany, 1971; Hamilton *et al.*, 1972; Lewis, 1975; Tovi and Mares, 1978).

Remnants of thymic tissue in the upper neck or the even larger mass of a complete ectopic thymus seem to be very rare (Rössle, 1932; Lewis, 1962; Barrick and O'Kell, 1969; Hinds *et al.*, 1970; Finch and Gough, 1972; Bhatnagar, 1974; Fahmy, 1974; Mikal, 1974; Shackelford and McAlister, 1974; Lewis, 1975; Maisel *et al.*, 1975; Johnsen and Bretlau, 1976; Tovi and Mares, 1978; Batsakis, 1979; Kaluzynski, 1980). Lewis, in 1962, evaluated 34 cases from the literature, one of which showed a lethal outcome due to obstruction of the trachea. Hinds *et al.*, in 1970, found 46 relevant cases in the English literature. Only 10% of these were solid thymus masses, with the rest showing already gross degeneration. 95% in Hinds's analysis were unilat-

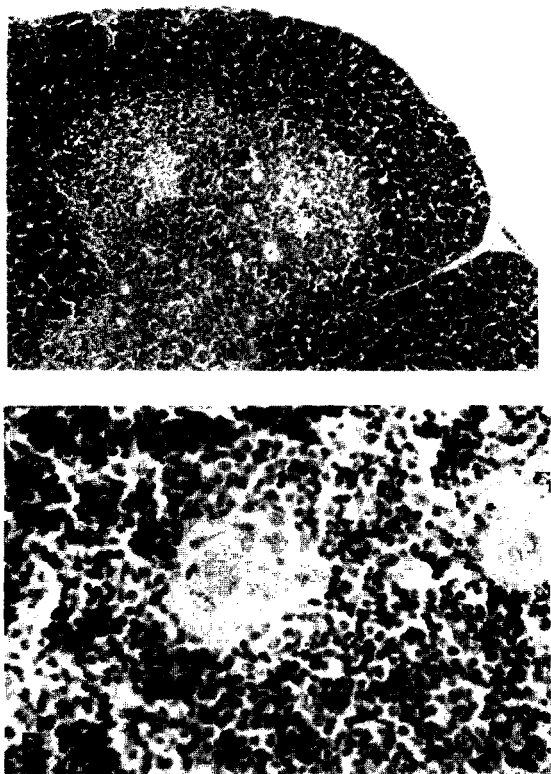


Fig. 4

Figure 4(A)—Regular thymic tissue with dark cortex and light medulla. Hematoxylin eosin, $\times 10$. (B) Two Hassall's corpuscles in the medulla. Hematoxylin eosin, $\times 70$

eral cases. The most comprehensive and more recent review of the literature revealed 68 cases of aberrant or ectopic thymus; 30 of these were normal tissue when removed and fall into the same category as our case (Tovi and Mares, 1978). The majority of the cases showed cystic degeneration and enlargement. Furthermore, neoplastic change into thymoma is not uncommon if we may relate 11 cases of this tumour in the neck described in a review in 1983 to the rarity of ectopic thymus in this area (Pirkey, 1954; Yamashita *et al.*, 1983).

Clinically undiscovered thymic tissue in the neck may, however, be more common than one would conclude from these figures. No relevant investigation exists of the neck in young children or adults. Maisel *et al.* (1975), in an examination of 67 fetuses, demonstrated that in the second half of the gestation period extensions of the thymus into the area between the thyroid and the mediastinum are still very common. In later life, however, thymus

tissue in this area was found in only 1.8% of 657 specimens from thyroid surgery (Yamashita *et al.*, 1983). Ectopic thymus in the lower neck is obviously far more common than in the upper neck, as in our case. Furthermore, there seems to be a considerable gap between post-mortem findings and clinically relevant cases.

The diagnosis of ectopic thymus has never been made preoperatively (Tovi and Mares, 1978). The differential diagnosis may include branchial, thyroglossal, thymic and parathyroid cysts, vascular tumour such as haemangioma, cervical hygroma, and lymphoma or soft solid neoplasms like lipoma. May, in 1976, presented an excellent and fairly comprehensive review of the differential diagnosis of such neck masses in children.

Because of the high incidence of pathological change, surgical removal of the ectopic thymus seems the treatment of choice (Tovi and Mares, 1978) and has been performed in all reported cases. Only Finch and Gough (1972) in their two cases replaced the extirpated tissue after frozen section diagnosis. Although the parathyroid glands derive from the same pharyngeal pouches as the thymus and in their earlier stage keep in close contact with the descending thymus, we found no such tissue in our case and only very few cases have been reported where parathyroid tissue was included in the removed ectopic thymus specimen (Lewis, 1962; Bhatnagar, 1974; Tovi and Mares, 1978; Kaluzynski, 1980). As the parathyroid anlage is bilateral, a simultaneous removal of the glands on one side with the ectopic thymus, though a rarity within rare cases, appears to be an acceptable consequence.

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