Medullary carcinoma

Medullary carcinoma

These are tumours of the parafollicular (C cells) derived from the neural crest that are not unlike those of a carcinoid tumour (Figure 55.26). High levels of serum calcitonin and carcinoembryonic antigen are produced. Calcitonin levels fall after resection and rise again with recurrence, making it a valuable tumour marker in the follow-up of patients with this disease. Diarrhoea is a feature in 30% of cases and this may - be due to 5-hydroxytryptamine or prostaglandins produced by the tumour cells. Medullary carcinoma may occur in combination with adrenal phaeochromocytoma and hyperparathyroidism - (usually due to hyperplasia) in the syndrome known as multiple endocrine neoplasia type 2A (MEN-2A). The familial form of the disease frequently a ff ects c hildren and young adults, whereas the sporadic cases occur at any age with no sex predominance. When the familial form is associated with prominent mucosal neuromas involving the lips, tongue and inner aspect of the eyelids, with a Marfanoid habitus, the syndrome is referred to as MEN type 2B (see Chapter 57). Involvement of lymph nodes occurs in 55–60% of cases and blood-borne metastases are common. Tumours are not TSH dependent and do not take up radioactive iodine. The prognosis is variable and depends on the stage at diagnosis. volvement virtually eliminates the prospect of Any nodal in cure and, unfortunately , even small tumours confined to the thyroid gland may have spread by the time of diagnosis, par ticularly in familial cancers. In common with many endocrine tumours the progression of disease may be very slow , with a characteristically indolent course and long survival, even in the . absence of cure In familial cases of medullary thyroid cancer, genetic screening of relatives should be recommended and the infor mation used to make recommendations concerning prophy . Some relatives may be monitored into lactic thyroidectomy adulthood with serial calcitonin monitoring. In contrast, the highest risk mutations are associated with early-onset disease and total thyroidectomy is recommended during infancy .

Figure 55.26 Histology of medullary carcinoma showing characteris tic ‘cell balls’ and amyloid (courtesy of Dr SWB Ewen, Aberdeen, UK).

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Revision #1
Created 2025-12-31 15:20:51 UTC by Omar Ayman
Updated 2025-12-31 15:20:51 UTC by Omar Ayman