PRIMARY TUMOURS OF THE NECK Neurogenic tumours

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Paraganglioma (carotid body tumour) This is a rare tumour that has a higher incidence in areas where people live at high altitudes because of chronic hypoxia leading to carotid body hyperplasia. The tumours most commonly present in the fifth decade. Approximately 10% of patients have a family history , with familial cases caused by mutations in the genes for succinate dehydrogenase (SDH) enzyme. There is an association with phaeochromocytoma in familial cases, and thus appropriate tests should be undertaken to rule out synchronous catecholamine-secreting tumours during the Charles Mantoux , 1877–1947, physician, Le Cannet, Alpes Maritimes, France, described the intradermal tuberculin skin test in 1908. Albert Leon Charles Calmette , 1863–1933, and Jean-Marie Camille Guérin the bacille Calmette–Guérin in 1908. ® -TB - - - work-up. The tumours arise from the chemoreceptor cells on the medial side of the carotid bulb and, at this point, the tumour is adherent to the carotid wall. These tumours are usually benign with only a small number of cases producing proven metastases (Figures 52.67 and 52.68). Clinical features There is often a long history of a slowly enlarging, painless lump at the carotid bifurcation. About one-third of patients present with a pharyngeal mass that pushes the tonsil medially and anteriorly . The mass is firm, rubbery , pulsatile, mobile from side to side but not up and down and can sometimes be emptied by firm pressure, after which it slowly refills in a pulsatile manner. A bruit may also be present. Swellings in the parapharyngeal space, which often displace the tonsil medially , should not be biopsied from within the mouth. Investigations When a paraganglioma is suspected, a carotid angiogram can be carried out to demonstrate the carotid bifurcation, which is usually splayed, and a blush, which outlines the tumour vessels. MRI scanning also provides excellent detail in most cases. This tumour must not be biopsied and fine-needle aspiration is also contraindicated. , 1872–1961, microbiologists at the Institute Pasteur, Lille, France, introduced

Ganglion nodosum Glomus jugulare Internal Glossopharyngeal jugular nerve vein Carotid bodies Vagus nerve Aorticopulmonary bodies Pulmonary artery Figure 52.67 Sites for chemodectomas.

Treatment The Shamblin classification is used to determine the surgical resectability of these tumours. Type I tumours are localised and do not involve more than 180° of the carotid vessels; type II tumours surround the vessel by over 180°; and type III tumours completely encase the vessels and are more challeng ing to resect with higher complications and a possible need for vessel reconstruction. Because these tumours rarely metastasise and their overall rate of growth is slow , the need for surgical r emoval must be considered carefully as complications of surgery are poten tially serious. The operation is best avoided in elderly patients. Radiotherapy will not cure the tumour but can prevent further growth. In some cases it may be possible to dissect the tumour away from the carotid bifurcation b ut, at times, when the tumour is large, it may not be separable

from the vessels and resection will be necessary , such that all appropriate facilities should be available to establish a bypass while a vein autograft is inserted to restore arterial continuity in the carotid system.

Figure 52.68 Axial view computed tomography angiogram (a) and magnetic resonance imaging

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