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Haemangiopericytoma of the supraclavicular region – multidetector computed tomography (MDCT) findings: a case report

Haemangiopericytomas (HPC) are rare vascular tumours arising from pericytes of Zimmermann, contractile spindle cells that surround the capillaries and post – capillary venules (4). HPC resembles about 1% of all vascular neoplasms and may occur anywhere in the body. It commonly arises in the retroperitoneum and lower extremities, with 15 to 30% occurring in the head and neck region (5). We report here a case of haemangiopericytoma of the right supraclavicular region, which is a very rare site of occurrence and the findings using multislice computed tomography.

CASE REPORT

A 44-year-old male was referred from Regional Oncology Centre with progressively increasing palpable mass in the right supraclavicular region. The CT examinations were performed with the 64-row LIGHTSPEED ULTRA (GE) in the pre-contrast scanning with slice thickness – 2.5 mm and post-contrast scanning – 1.25 mm thickness. The contrast medium in the amount of 100–120 ml was injected with the automatic power injector and speed of 4 ml/s. The scanning delay was determined on the basis of the density measurements using the Smart Prep technique. Multiphasic scanning protocol was applied, enabling visualization of the tumour in the arterial and venous phase. Postprocessing was performed using the Advantage 4.2 workstation (GE).

On the first examination (March 2006) non-enhanced scans showed a soft tissue mass located in the right supraclavicular region, adjacent to right jugular vein and the right pleura apex. The lesion was irregular in shape, measuring 51x45x50 mm, with the small foci of calcification in its central part. On native scans the central part of the tumour revealed density of 8 Hounsfield units and in the peripherals – approximately 32 HU. On contrast medium administration, in the arterial phase, the peripherals' density increased up to 90 HU, with the central part density about the same limits – suggesting central necrosis. Also, feeding vessels from thyroid artery and external carotid artery branches were depictured. Additionally, in the right parapharyngeal space, a small lesion (25x15 mm in size), with similar density and contrast enhancement was visualized.

The next two examinations (April 2006; June 2006) demonstrated gradual decreasing in size of the tumour and the lesion in the right parapharyngeal space, with no evident changes in post-contrast enhancement pattern.

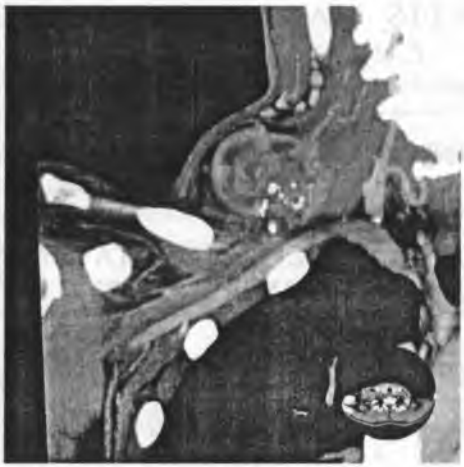


Fig. 1. A CT scan – oblique MIP reconstruction showing highly vascular tumour with small focal calcifications

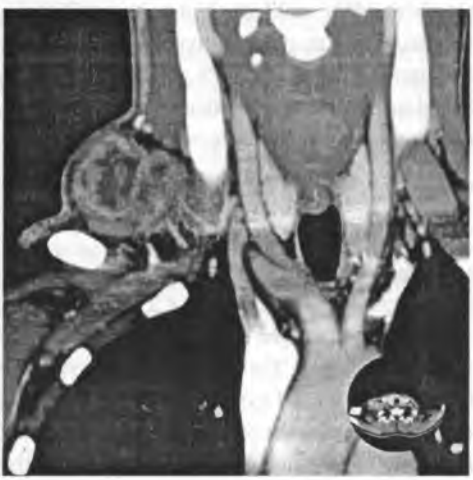


Fig. 2. A MIP reconstruction depicting small feeding vessels from thyroid artery and external carotid artery branches

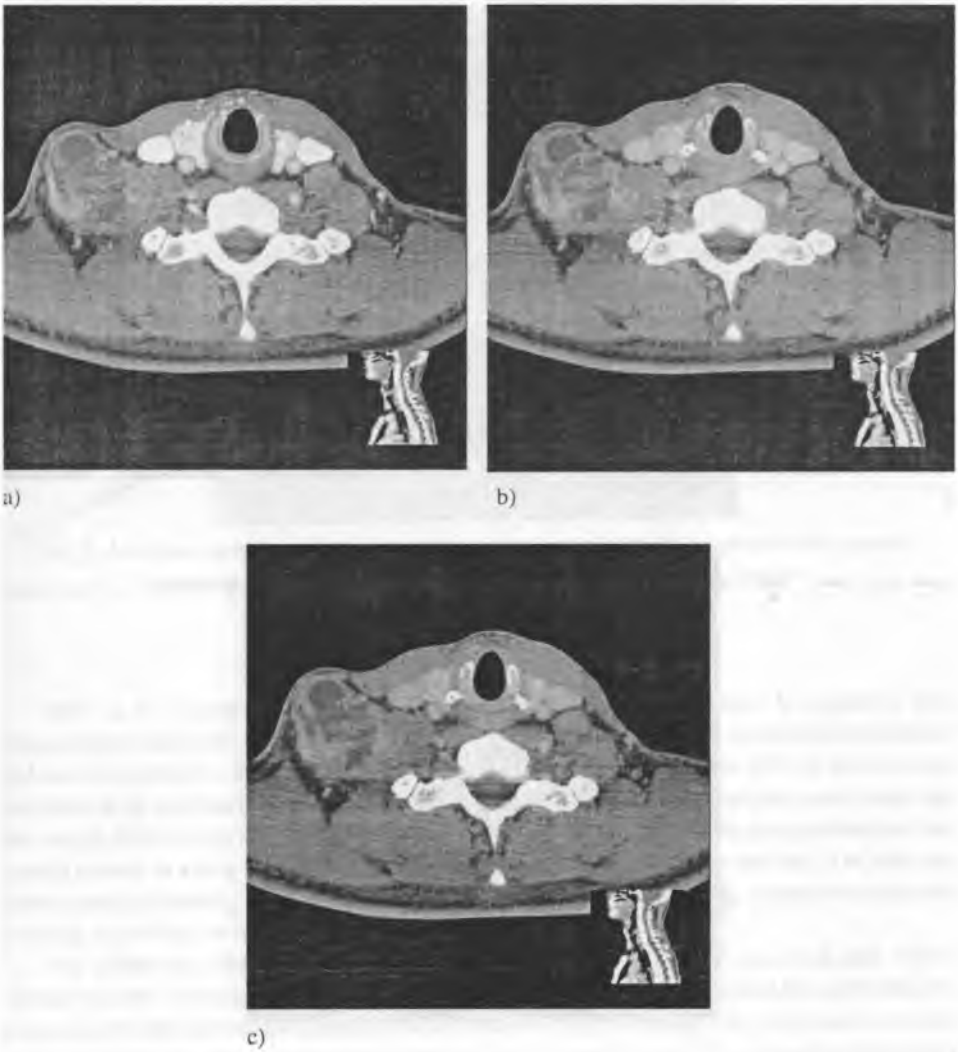
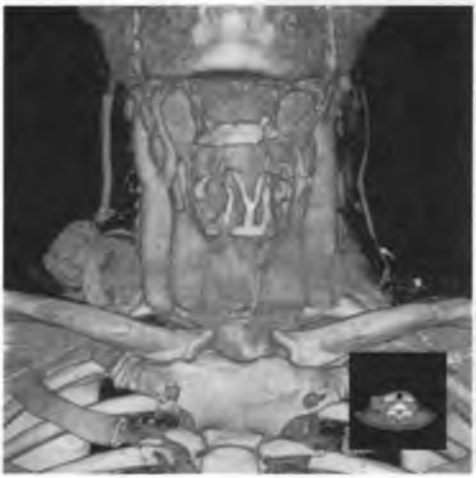
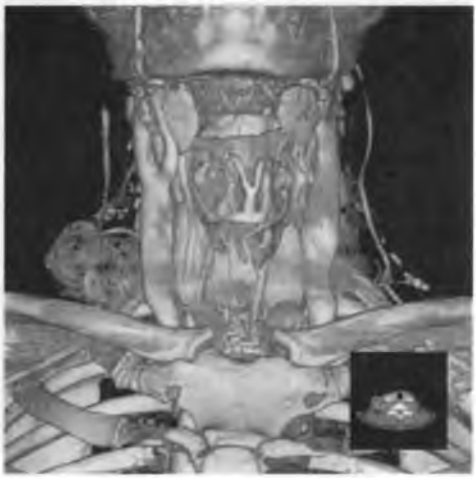


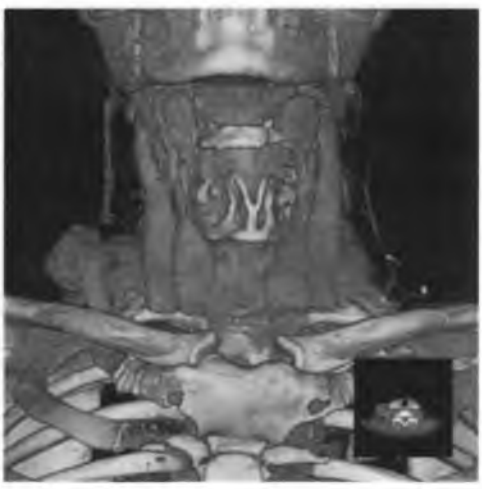
Fig. 3. CT scans, an axial view in: a) arterial phase, b) parenchymal phase, c) delayed phase



a)



b)



c)

Fig. 4. A CT scans – Volume Rendering reconstructions in: a) arterial phase, b) parenchymal phase, c) delayed phase

Fig. 5. A 3-dimentional reconstruction with the maximum intensity focused on skin density, showing deformation of the skin in the right supraclavicular region

DISCUSSION

HPC is an uncommon vascular tumour that is difficult to diagnose. It originates from Zimmermann's pericytes (5) – a line of cells that is supposed to be responsible for blood flow regulation and vessel contraction. It may arise in any part of the body, and from 15 to 30% of these tumours are found in the head and neck region. It affects males and females equally and can occur at any age, but usually (80%) arises after 2nd decade (5). Haemangiopericytomas have no race predilection, they usually present as a slow-growing mass, that is often nodular and well circumscribed. The overlying skin or mucosa is usually normal in appearance. The tumour therefore usually presents very late after reaching a size large enough to produce pressure effect.

The differential diagnosis includes lymphomas, neurofibromas, fibrosarcomas and solitary fibrous tumours. Imaging, biopsy and immunohistochemical staining are essential for establishing the diagnosis (3). On microscopic examination the cells are spindle-shaped with cytoplasmic extension and indistinct border. The eccentrically located nucleus is round or oval (4). Assessed on histological (cellularity, nuclear atypia and mitotic activity) and clinical basis (hemorrhage, necrosis and tumour size) haemangiopericytomas are classified as benign, borderline and malignant (5).

The patient presents with a slowly increasing mass in the right supraclavicular region and symptoms being mostly due to pressure on adjacent structures. On CT examination it appears as a soft tissue density mass lesion with heterogeneity due to foci of necrosis, hemorrhage and cystic degeneration as well as small focal calcifications; with intense heterogeneous enhacement on contrast medium administration especially in peripheral part of the tumour. The MIP – reconstruction images demonstrate vascular nature of the tumour, reveal the exact source of the blood supply – mostly from thyroid arteries and external carotid artery branches. The examination reveals the presence of additional, pathological structure (app. 25x12 mm in size) in the right parapharyngeal space. The similar structure and post-contrast enhancement suggests the metastatic nature of the lesion. Because of the rarity and unpredictable biologic behaviour of these neoplasms, there is controversy about the best way to manage them. It is generally accepted that the treatment of choice is radical surgical

excision when the tumour is localized and technically respectable. The combination of radiation therapy and chemotherapy is recommended in cases of unresectable lesions and in presence of metastatic spread of the disease (3). In all cases the long-term follow-up is essential in order to evaluate the treatment response of the tumour as well as due to high incidence of local recurrence (after surgical resection).

Multidetector computed tomography (MDCT) evaluation of pathological masses allows data to be acquired during optimal enhancement, which aids in tumour visualization and differentiation. This technology permits thinner slices to be acquired during multiphase scanning, with improved spatial resolution. The use of multiplanar reformatted images and 3-dimensional representations of the vascular structures helps in accurate diagnosing and aids in successful surgical resection (if possible).

Enzinger and Smith (1) evaluated 106 haemangiopericytomas and concluded that the commonest site is lower extremity (35%) followed by pelvis or retroperitoneum (25%), head and neck (16%), trunk (14%) and upper extremity (10%). However, there has been no report of this kind of tumour in the supraclavicular region. Additionally, no data can be found in the literature, regarding appliance of multidetector computed tomography (MDCT) with multiphase scanning protocol.

Hamenagiopericytoma should be considered in the differential diagnosis of any highly vascular tumour anywhere in the body. Multidetector computed tomography with contrast enhancement, especially with multiphase scanning protocol, is a valuable tool in diagnosis, evaluation of treatment as well as the follow-up in patient with haemangiopericytomas.

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SUMMARY

Haemangiopericytoma is a rare, highly vascular tumour, that is thought to originate from the vascular pericytes of Zimmermann. We report a case of a 44-year-old man with a haemangiopericytoma located in the right supraclavicular region. We present an application of multidetector computed tomography (MDCT) with multiphase scanning protocol in evaluation of the primary tumour, additional lesions and post-treatment follow-up. A brief review of the pathology and nature of the tumour is provided.

Haemangiopericytoma okolicy nadobojczykowej prawej w obrazowaniu wielorzędowej tomografii komputerowej (MDCT) – opis przypadku

Haemangiopericytoma jest rzadko spotykanym, bogato unaczynionym guzem, wywodzącym się najprawdopodobniej z pericytów Zimmermanna. Prezentujemy przypadek 44-letniego mężczyzny z guzem typu haemangiopericytoma, zlokalizowanym w okolicy nadobojczykowej prawej. Przedstawiamy obrazowanie przy pomocy wielorzędowej tomografii komputerowej (MDCT) w protokole wielofazowym, jej zastosowanie w dokładnej ocenie guza, zmian dodatkowych jak również w ocenie pooperacyjnej. Podajemy również skrócone informacje dotyczące opisywanego typu nowotworu.