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Neuroendocrine tumour metastasis to the orbit

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Abstract

Neuroendocrine neoplasms are tumours that usually arise in the gastrointestinal tract and the bronchopulmonary system. The orbit is a rare anatomical site for their metastases. In the following article we present a case report of a 73-year-old man who was admitted to the Ophthalmology Department because of eye pain and high intraocular pressure in the right eye. There was also eye motility restriction, diplopia, oedema of the eyelid, and subconjunctival haemorrhage. Magnetic resonance imaging revealed a tumour in the right orbit, probably arising from the inferior rectus muscle. The patient was qualified for surgery, during which orbit decompression was conducted and a sample of the tumour tissue was collected. Based on the biopsy of the lesion, diagnosis of a metastatic neuroendocrine neoplasm, probably of gastrointestinal origin, was made. Further diagnostic procedures revealed metastases to other organs, and the patient was qualified for oncological treatment. In this case, orbital metastasis was the first diagnosed location of the neoplasm. **(Endokrynol Pol 2019; 70 (5): 455–456)**

Key words: *neuroendocrine neoplasm; carcinoma; orbit; extraocular muscles; metastasis; intraocular pressure*

Case report

A 73-year-old man was admitted to the Department of Ophthalmology because of pain and high intraocular pressure (45 mm Hg) in the right eye. The patient also complained about pain in the epigastrium and abdomen, weakness, vomiting, trembling hands, fever, weight loss of about 15 kilograms in the past 1–1.5 months, and increasing memory problems in the last two years. Right eye movements were restricted in all directions, especially vertical, diplopia while looking to the far left, left-down, down, right-down, up-right, up, proptosis of the right eyeball, and hypertropia. In addition, eyelid oedema and subconjunctival haemorrhage were noted. Systemic and topical intraocular pressure-lowering drugs and methylprednisolone intravenously were administered. Afterwards, the patient reported good well-being and reduced eye pain, but diplopia was still present.

Magnetic resonance imaging of the orbits was conducted. An extraocular tumour of the right orbit was revealed, probably arising from inferior rectus muscle, with features of haemorrhage to the tumour. The dimensions of the lesion were 19 mm × 20 mm × 17 mm. It was heterogeneous, with strong marginal intensification (Fig. 1).

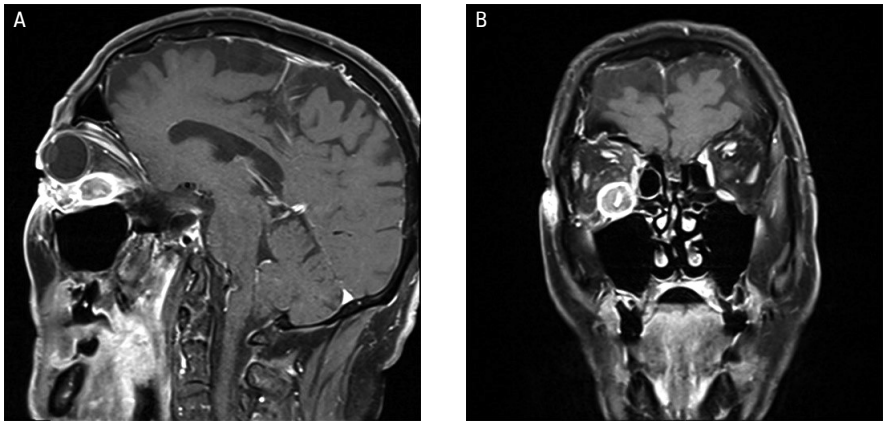

Figure 1. *Magnetic resonance imaging of the orbits.* **A.** *Sagittal view of the extraocular tumour in the right orbit.* **B.** *Coronal view of the extraocular tumour in the right orbit*

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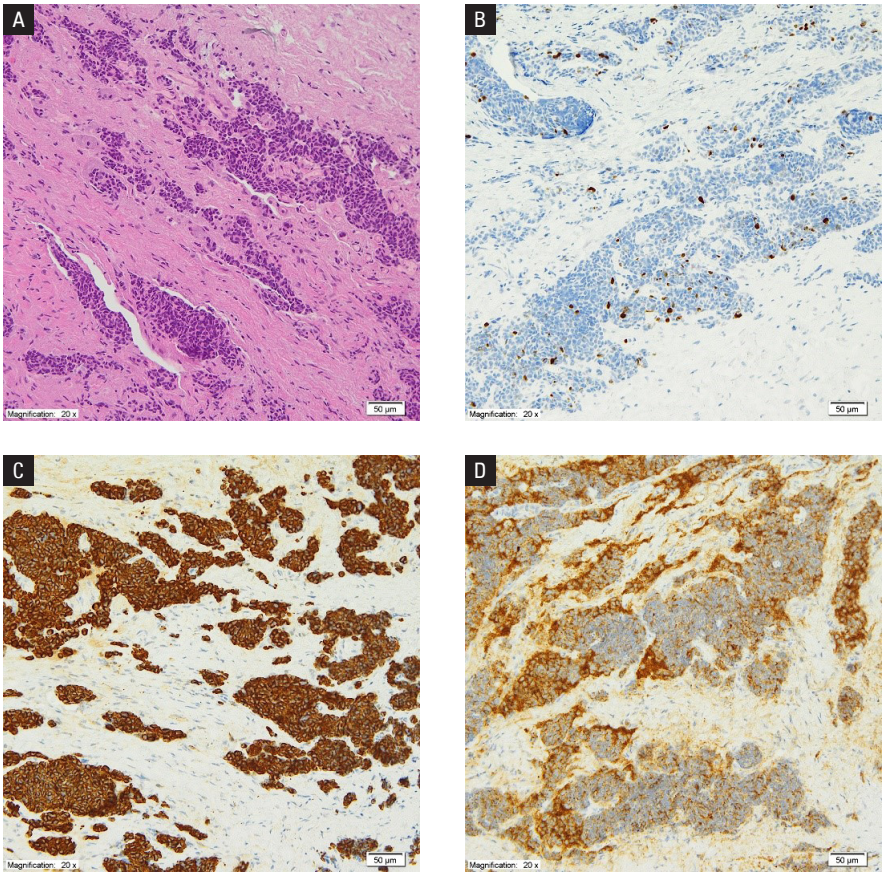

Figure 2. *Histology with immunohistochemistry of the neuroendocrine tumour metastasis to the extraocular muscle.* **A.** *Haematoxylin and eosin staining.* **B.** *Ki-67 staining.* **C.** *Cytokeratin staining.* **D.** *Synaptophysin staining*

The patient was qualified for surgery aimed at orbit decompression and sampling of the tumour so as to introduce further therapy. The process of orbit decompression was managed by two approaches, without proceeding to functional endoscopic sinus surgery. After nasal mucous membrane anemisation, the middle concha was removed. The ethmoid and maxillary sinuses were opened. The medial part of the upper wall of the maxillary sinus (medially from canal of the maxillary nerve) and medial (ethmoidal) wall of the orbit were removed, exposing only fat tissue, which was taken for histopathological examination. After separation of the eyeball conjunctiva inferior rectus muscle was grasped. The tumour tissue was tough, solid within the muscle fibres, whereas the eyeball was not infiltrated. A sample of the tissues surrounding the muscle was also collected for histopathological examination.

Analysis of the tumour sample confirmed a neuroendocrine neoplasm with low mitotic index (1/10 HPF) and immunophenotype: CKAE1/AE3+, CK7–, CK20–, TT-F1–, SMAD4+, CDX2+, PSA–, synaptophysin+, and Ki67 index of 3–5%. The result mostly resembled gastrointestinal neuroendocrine neoplasm metastasis (Fig. 2).

Abdominal ultrasonography and computed tomography revealed scattered, multiple secondary lesions in the liver, multifocal pathological lesions in the pancreas (with suspicion of being the origin of the neoplasm), and enlarged abdominal lymph nodes. Furthermore, positive scintigraphy of somatostatin receptors was detected. The patient was qualified for somatostatin analogue treatment.

Three weeks after surgery, a follow-up examination revealed right eye intraocular pressure of 9.1 mm Hg (without taking any intraocular pressure lowering drugs), slight right eye proptosis, residual diplopia, and slight right eye movement restriction to the right and up. In another follow-up examination about 3.5 months after surgery, the patient's intraocular pressure was normal, whereas diplopia persisted in all directions, except for to the left and straight ahead.