Giant somatotroph adenoma

Patients with acromegaly usually harbor pituitary macroadenomas measuring between 10 and 30 mm in maximal diameter. Giant (adenoma size ≥40 mm) GH secreting pituitary neuroendocrine tumor are rarely encountered.

They are invasive, uncontrolled by surgery, and respond poorly to medical treatment. Aggressive multimodal therapy is critical for their management, enhancing control rate and biochemical remission [1].

Medical therapies for GHPAs, including somatostatin analogs and GH receptor antagonists, are becoming increasingly important adjuncts to surgical intervention. Stereotactic radiosurgery serves as an important fallback therapy for tumors that cannot be cured with surgery and medications. Data suggest that patients with aggressive and refractory GHPAs are best treated at dedicated tertiary pituitary centers with multidisciplinary teams of neuroendocrinologists, neurosurgeons, radiation oncologists and other specialists who routinely provide advanced care to GHPA patients. Future research will help clarify the defining features of “aggressive” and “atypical” PAs, likely based on tumor behavior, preoperative imaging characteristics, histopathological characteristics, and molecular markers [2].

Case series

2017

Giordano et al., present the clinical, radiological and hormonal status of three patients affected by invasive GH-secreting pituitary neuroendocrine tumors without clinical signs and symptoms of acromegaly with elevation of serum IGF-1 from a series of 142 pituitary neuroendocrine tumors operated in the Department of Neurosurgery, International Neuroscience Institute-Hannover, Germany with the aid of intraoperative magnetic resonance imaging (MRI). Total tumor removal was possible in two of the three cases; the patients show normal hormonal status and no recurrence at long-term follow-up. In the third case, due to the different features of the tumor, complete resection was not possible and a multimodal treatment was performed that allowed regularization of the hormonal status and control of the residual tumor. GH-secreting adenomas without clinical manifestation of acromegaly are uncommon lesions. Total microsurgical excision can be curative. However, in case of partial removal, a tailored adjuvant treatment should be considered to preserve the quality of life of the patient and avoid regrowth of the lesion. In not resectable tumors, preoperative medical treatment with somatostatin analogues is always an option [3].

2015

Shimon et al. identified 34 patients (15 men and 19 females) with giant adenomas among 762 subjects (4.5%) with acromegaly, and characterized their clinical characteristics and response to treatment.

Mean age at diagnosis was 34.9±12.5 years (range, 16-67 years). Mean adenoma size was 49.4±9.4mm (range, 40-80mm); 30 adenomas showed cavernous sinus invasion and 32 had suprasellar extension. Twenty-nine (85%) patients had visual field defects. Mean baseline IGF1 was 3.4±1.8×ULN. All patients except one underwent pituitary surgery (one to three procedures), but none achieved hormonal remission following first surgery. Among the 28 subjects with visual disturbances, 14 recovered post-operatively and 13 improved. Treatment with somatostatin analogs was given to all patients after surgical failure. Six achieved remission, nine others were partially controlled (IGF1<1.5×ULN; 3/9 when combined with cabergoline), and 17 did not respond (two were lost). Nine patients were treated with pegvisomant, alone (n=4) or in combination with somatostatin analogs (n=5); five are in remission and two are partially controlled. Pasireotide-LAR achieved hormonal remission in one of the six patients. Currently, after a mean follow-up period of 8.9 years, 17 patients are in biochemical remission, eight are partially controlled, and seven are uncontrolled (two were lost to follow-up).

Giant GH-secreting adenomas are invasive, uncontrolled by surgery, and respond poorly to medical treatment. Aggressive multimodal therapy is critical for their management, enhancing control rate and biochemical remission [4].

Case reports

A 23-year-old male patient presented with continuous increase in height during the past 6 years due to a GH-secreting giant pituitary neuroendocrine tumor. Because of major intracranial extension and failure of octreotide treatment to shrink the tumour, the tumour was partially resected by a trans-frontal surgical approach. At immunohistochemistry, the tumour showed a marked expression of GH and a sparsely focal expression of prolactin. Somatostatin receptors (sst) 1-5 were not detected. Tumour tissue weakly expressed dopamine receptor type 2. The Gs alpha subunit was intact. Conversion from somatostatin analogue to pegvisomant normalized insulin-like-growth-factor-I (IGF-I) levels and markedly improved glucose tolerance.

Pegvisomant is a potent treatment option in patients with pituitary gigantism. In patients who do not respond to somatostatin analogues, knowledge of the SST receptor status may shorten the time to initiation of pegvisomant treatment [5].

1)
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