Restrictive extraocular myopathy: A presenting feature of acromegaly

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A 45-year-old man presented with binocular diplopia in primary gaze for 1 year. Orthoptic evaluation showed 10-prism diopter right eye hypotropia and 6-prism diopter right eye esotropia. The elevation and abduction of the right eye were mechanically restricted. This was associated with systemic features suggestive of acromegaly. Magnetic resonance imaging (MRI) of the brain demonstrated a pituitary macroadenoma. An elevated serum insulin-like growth factor I level and the failure of growth hormone suppression after an oral glucose load biochemically confirmed the diagnosis of acromegaly. Computed tomography (CT) of the orbit demonstrated bilateral symmetrical enlargement of the medial rectus and inferior rectus muscle bellies. All tests regarding Graves-Basedow disease were negative. Although rare, diplopia due to a restrictive extraocular myopathy could be the presenting symptom of acromegaly.

Key words: Acromegaly, diplopia, extraocular myopathy, pituitary adenoma, restrictive myopathy

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The incidence of acromegaly is approximately three cases per one million persons per year and more than 90% of patients with acromegaly have a growth hormone (GH) secreting pituitary adenoma. GH induces the synthesis of insulin-like growth factor I (IGF-I). The clinical manifestations are attributable to high serum concentrations of both GH and IGF-I. They range from subtle signs of acral overgrowth, soft-tissue swelling, arthralgias and jaw prognathism to florid osteoarthritis, diabetes mellitus and respiratory and cardiac failure. In addition, the adenoma may cause local symptoms such as headache, visual field defects and cranial nerve palsies.[1]

Extraocular muscle enlargement has been associated with acromegaly[2-4] and may rarely be the presenting feature of the disease.[5,6] We report a patient with diplopia, due to restrictive extraocular myopathy, as the presenting symptom of acromegaly. To our knowledge, this finding has not been reported previously.

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Case Report

A 45-year-old man presented with binocular diplopia in primary gaze for 1 year without any other ocular or general complaints. Past medical history was unremarkable. Uncorrected visual acuity was 20/20 in both eyes. Pupillary reflexes were normal, slit-lamp examination was unremarkable and there was no proptosis (Hertel exophthalmometry readings: 11 mm both eyes), ptosis or eyelid swelling. Fundoscopy and Goldmann kinetic perimetry were normal. Orthoptic evaluation showed 10-prism diopter right eye hypotropia and 6-prism diopter right eye esotropia on cover test for near fixation. The elevation and abduction of the right eye were clinically restricted. Hess charting of the right eye showed restricted elevation in abduction (12°) and restricted abduction (5°). Hess charting of the left eye showed increased elevation in adduction (9°) and increased adduction (4°) [Fig. 1]. The forced duction test was positive. Computed tomographic (CT) scan of the orbit demonstrated bilateral symmetrical enlargement of the medial rectus and inferior rectus muscle bellies [Fig. 2]. Physical examination revealed enlarged hands, coarse facial features and macrognathia [Fig. 3]. Diagnosis of acromegaly was suspected and magnetic resonance imaging (MRI) was performed, demonstrating a pituitary macroadenoma (12 mm) within a deformed sella turcica [Fig. 4]. Suprasellar cistern or optic tract was not involved. An elevated serum IGF-I level and the failure of GH suppression after an oral glucose load biochemically confirmed the diagnosis of acromegaly. Endocrinological work-up also revealed an associated non–insulin-dependent diabetes mellitus. Thyroid gland was clinically enlarged and ultrasonography demonstrated a multinodular goiter. Repeated thyroid function tests (thyroid-stimulating hormone, free triiodothyronine, free thyroxine) were normal. Thyroid peroxidase antibody, thyroglobulin antibody and thyroid-stimulating hormone receptor antibody levels were normal. The patient was treated first with octreotide, a somatostatin analogue. Later on, he underwent transsphenoidal surgical resection of the pituitary macroadenoma. The results of the cover test and Hess screen test remained unchanged during a 2-year follow-up.

Discussion

The clinical features of acromegaly develop insidiously over decades, often resulting in a delay of 7–10 years in diagnosis after the estimated onset of symptoms.[1] Early recognition is essential as effective therapies can prevent disease progression and return the lifespan to normal.[7] Decreased vision or visual field defects are the main presenting complaints that lead to the work-up and diagnosis of acromegaly in 5.7% of the patients.[7] Diplopia could be due to cranial nerve palsy or hemifield slide phenomenon.[8]

In our case, binocular diplopia was the presenting complaint that brought the patient to medical attention. There were no cranial nerve palsies or visual field defects. The diplopia was caused by the bilateral enlargement of the medial rectus and inferior rectus muscles and increased on ductions away from the mechanically restricted muscles. The extraocular muscles

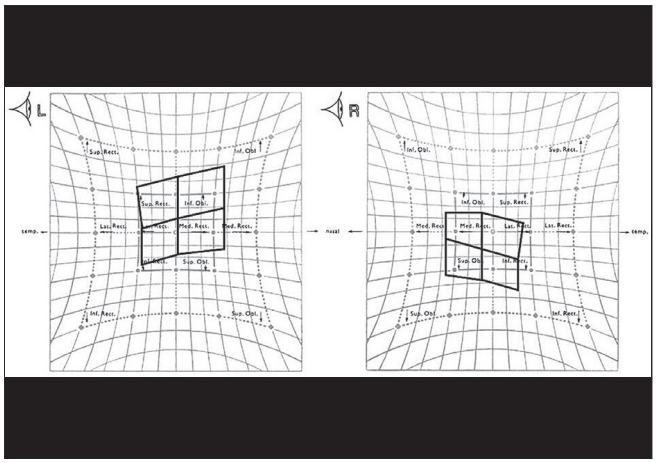

Figure 1: Hess screen test

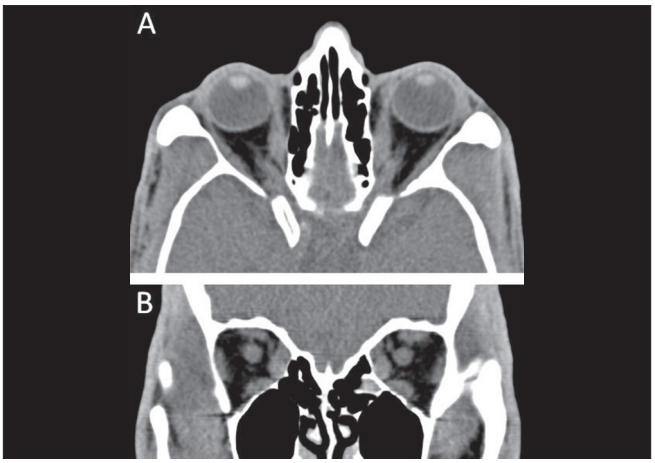

Figure 2: (A and B) Axial and coronal CT scans demonstrate bilateral enlargement of the medial rectus and inferior rectus muscle bellies

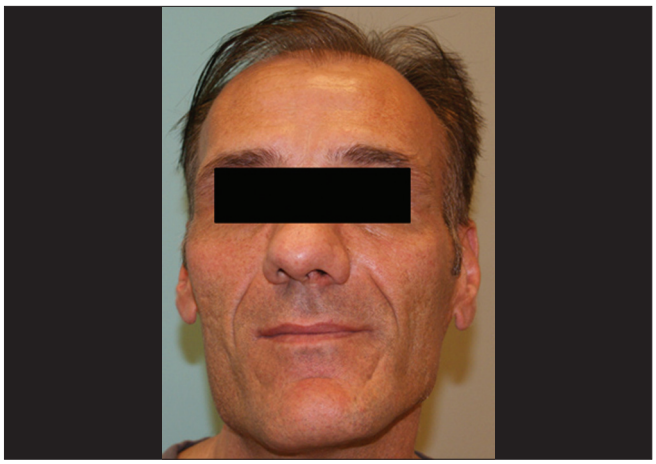

Figure 3: Coarse facial features and macrognathia

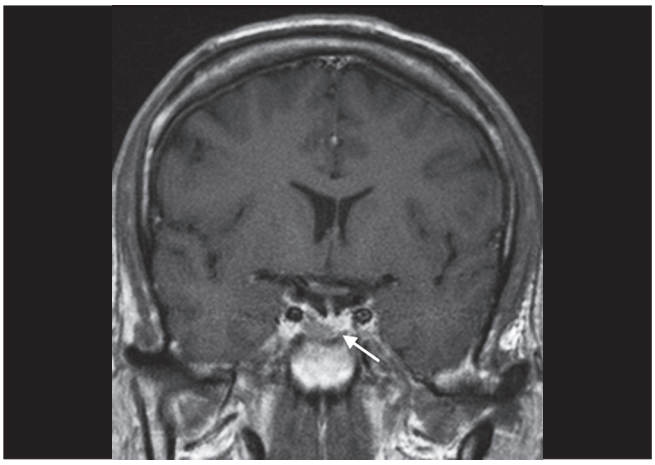

Figure 4: MRI scan with gadolinium illustrates a pituitary macroadenoma (arrow) within a deformed sella turcica

may be involved as part of the generalized organomegaly associated with acromegaly.[3,4] On CT scan, the muscles are diffusely and symmetrically enlarged. The degree of enlargement is related to duration of disease rather than hormone level.[2] Muscle biopsy specimens of the quadriceps muscle in acromegaly showed a hypertrophy of type 1 fibers and atrophy of type 2 fibers. Hypertrophy is probably a direct result of excess GH secretion.[9]

A PubMed search revealed two cases that have been reported previously with extraocular muscle enlargement as the presenting feature of acromegaly in combination with bilateral lower eyelid swelling[6] or ptosis.[5] Clinically significant motility defects were previously unrecognized in patients with GH-secreting pituitary tumors.[2]

Extensive endocrinological work-up was performed and all tests were negative regarding Graves-Basedow disease, the most common cause of extraocular muscle enlargement.[2-4] Although we cannot entirely exclude this disease, repeated laboratory testing during a 2-year follow-up was carried out and remained negative. Symptoms of thyrotoxicosis were absent. GH and IGF-I excess causes thyroid overgrowth, which is usually considered a typical aspect of the organomegaly developing in acromegaly. A goiter is revealed in 25–92% of cases. Multinodular goiter is detected in 65% of patients and nodular goiter is toxic in 14% of patients.[10] The most common nonthyroid causes of extraocular muscle enlargement are inflammatory, vascular, neoplastic and infectious processes.[2] Based on clinical presentation and CT scan of the orbit, the existence of these disorders is very unlikely.

To our knowledge, this is the first report of a patient with acromegaly with binocular diplopia due to restrictive extraocular myopathy as the initial clinical complaint.

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