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Focal Orbital Amyloidosis Presenting as Rectus Muscle Enlargement: CT and MR Findings

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Summary: We report a case of focal orbital amyloidosis involving rectus muscles, which is an extremely rare clinical condition. CT scans showed rectus muscle enlargement with punctate calcifications. Heterogeneous hypointensity was present on T2-weighted MR images, and homogeneous enhancement was seen on fat-saturated contrast-enhanced images of the muscles. These imaging findings seem to be suggestive of amyloidosis. Focal amyloidosis should be included in the differential diagnosis of extraocular muscle enlargement.

Clinically, amyloidosis is categorized into two main forms, systemic and localized. Systemic amyloidosis is a serious and usually fatal condition in which accumulation of amyloid fibrils in the tissues destroys normal structure and function. On the other hand, the localized form of amyloidosis is extremely rare, frequently involves the head and neck without systemic manifestations, and carries an excellent prognosis (1, 2). Although the CT appearance of orbital amyloidosis had been reported in some cases (1, 3–12), the MR findings of orbital amyloidosis have rarely been described. We report a case of orbital amyloidosis presenting as rectus muscle enlargement. CT scans showed marked enlargement of the rectus muscles with punctate calcifications. T2-weighted MR images showed heterogeneously hypointense signal in the muscles. The muscles enhanced markedly and homogeneously on contrast-enhanced MR images with fat saturation. These imaging appearances might be suggestive of amyloidosis, even though focal amyloidosis strictly localized in the extraocular muscles is extremely rare (8–11).

Case Report

A 47-year-old man had a 4-year history of slowly progressive double vision and left exophthalmos without accompanying pain. Prednisolone (40 mg daily for 3 months) was not effective. The patient was otherwise well, and there was no family history of amyloidosis. On admission, physical examination was negative except for mild left exophthalmos and restricted movement of the left eye in all directions. The patient had normal visual

acuity and fundi. Intraocular pressure was 15 mm Hg bilaterally. Blood and urine laboratory findings were normal. There was no clinical and laboratory evidence of Graves disease. Chest radiographs and ECG were also normal. Orbital CT scans showed marked fusiform enlargement of the left inferior and medial rectus muscles with sharp borders and punctate calcifications in the inferior rectus muscle (Fig 1A). The left lateral rectus muscle was mildly enlarged. The tendon of each enlarged muscle was spared. The signal intensity of the enlarged muscles was normal on T1-weighted MR images (Fig 1B) and was heterogeneously hypointense on T2-weighted images relative to the right inferior rectus muscle (Fig 1C). These muscles enhanced markedly after intravenous injection of gadopentetate dimeglumine (Fig 1D). Paranasal sinuses were normal. A biopsy with anterior septal approach was performed and a specimen was obtained from the margin of the left inferior rectus muscle. Microscopic examination of the specimen revealed extracellular amorphous and eosinophilic hyaline material (Fig 1E), which stained pink with Congo red (Fig 1F) and showed characteristic green birefringence when viewed under polarized light (Fig 1G). The specimen was decolorized after treatment with potassium permanganate. The diagnosis of amyloidosis (AA form) was made histopathologically. Rectal biopsy performed after surgery was negative for amyloid. No systemic amyloidosis or chronic inflammatory disease was apparent.

Discussion

The localized form of amyloidosis is extremely rare (1–20). Only 4% of focal amyloidosis involving the head and neck region occur in the orbit (1). In most cases of focal orbital amyloidosis, amyloid deposits are found in the eyelid or conjunctiva and in the superior portion of the orbit (3–7, 10, 12, 18–22). The typical clinical picture is of a painless palpable mass or exophthalmos present for years (3, 5–10, 12, 13, 20). Ptosis, double vision, and periorbital hemorrhage may occur (3–12, 18, 19, 21).

The CT appearance of orbital amyloidosis has been reported in some cases. Localized amyloidosis of the lacrimal gland (4, 5, 7, 10, 12, 20) and involvement of extraocular muscle have been demonstrated (1, 6, 7, 10, 12). Adjacent bone changes are seen in a few cases; erosion or focal thinning (7, 12) and hyperos-

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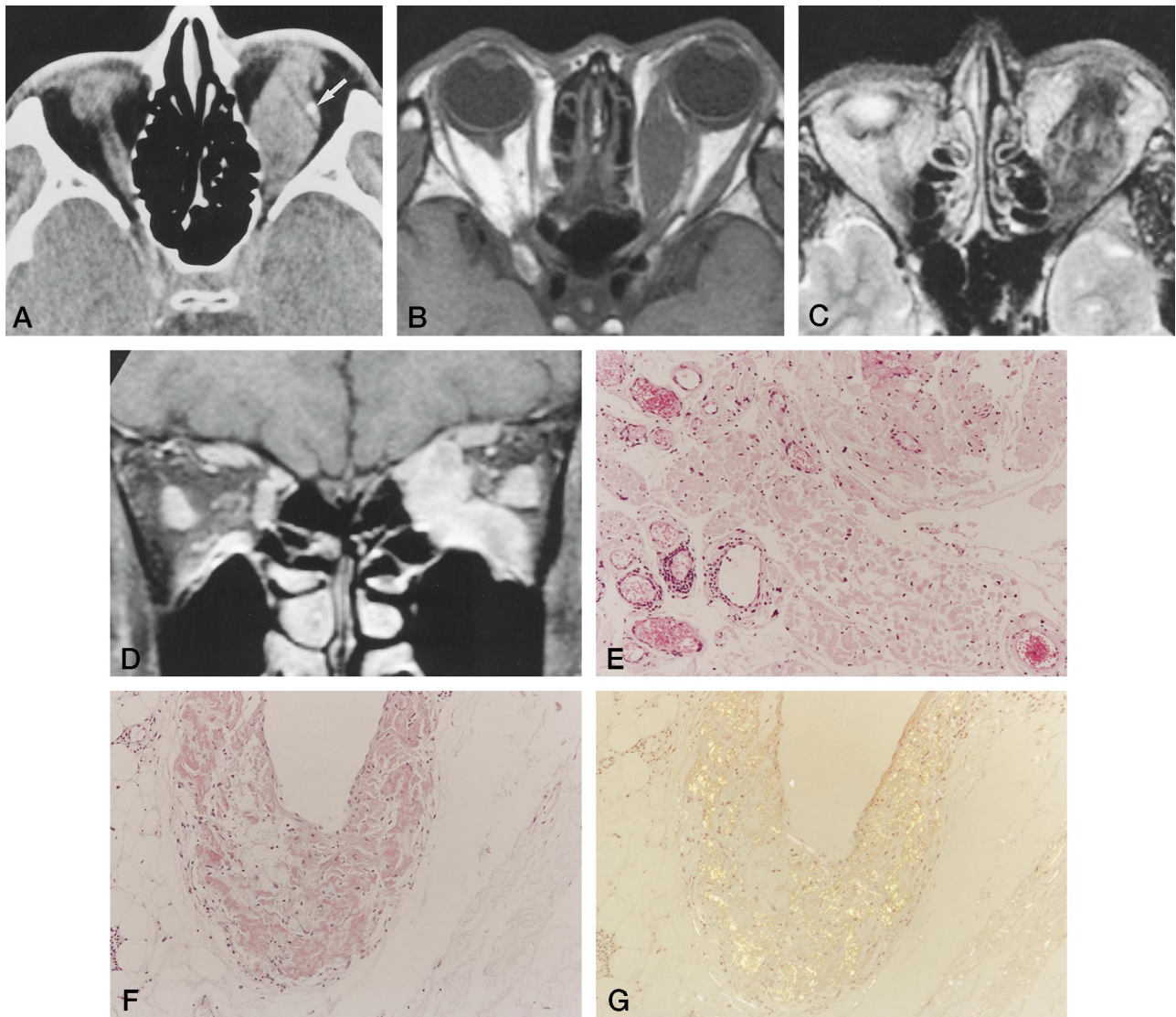


FIG 1. 47-year-old man with orbital amyloidosis involving the rectus muscles.

A, CT scan shows marked swelling of the left inferior rectus muscle with punctate calcification (arrow).

B-D, Spin-echo (SE) and fast spin-echo (FSE) MR images show marked enlargement of the left inferior and medial rectus muscles and mild swelling of the left lateral rectus muscle. Signal intensity of the enlarged muscles is normal on T1-weighted image (B) (SE, 600/15/2) and is heterogeneously decreased on T2-weighted image (C) (FSE, 3000/90/2). The enlarged muscles show marked enhancement on fat-saturated T1-weighted image obtained after contrast injection (D). Paranasal sinuses are normal.

E-G, Biopsy specimen obtained from the margin of the left inferior rectus muscle. Amorphous and eosinophilic material is seen extracellularly in the specimen (hematoxylin-eosin, original magnification $\times 250$) (E). The material is stained pink with Congo red (F) and shows characteristic green birefringence with polarized light (G).

tosis or focal thickening (12) have been reported. Punctate calcifications have been observed in seven of 16 cases of orbital amyloidosis (1, 4, 6, 7, 12) and in five of six cases of amyloidosis in other regions of the head and neck (1, 13, 16, 17). Although extraocular muscle enlargement can result from a wide variety of disease processes (12-27), strictly localized amyloidosis in the extraocular muscle is rare (5, 8, 9, 11). Accompanying calcification with enlarged extraocular muscles is rarely noted. In the head and neck region, enhancement of the amyloid lesion after injection of contrast material varies from none to marked on CT scans (1, 3, 4, 6, 7).

MR imaging of focal amyloidosis has rarely been reported. Hypointense signal has been observed in

the lesion on T2-weighted images in a patient with focal nasopharyngeal amyloidosis (1). Amyloid deposition in other organs also appears hypointense on T2-weighted images (28-30). Similarly, the lesion in our case was hypointense on T2-weighted images. Signal intensity of the enlarged extraocular muscles on T2-weighted MR images is variable in pathologic conditions: the signal is increased in Graves disease (31) and metastatic breast carcinomas (32); isointense or hyperintense to fat in orbital pseudotumor, infectious myositis, and sarcoid (32); and normal in congenitally enlarged extraocular muscles (27). Some granulomatous diseases or metastatic tumors may show hypointensity in the enlarged extraocular muscles without contiguous and/or adjacent lesions. In

addition, the combination of calcification on CT and heterogeneous hypointensity on T2-weighted MR imaging may be seen in orbital cavernous hemangiomas. However, this constellation might suggest focal amyloidosis as well.

Conclusion

When the imaging findings described in this report are seen in enlarged extraocular muscles, orbital amyloidosis should be included in the differential diagnosis, even though extraocular muscle enlargement is an extremely rare imaging manifestation of focal amyloidosis.

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