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Midline granuloma presenting as orbital cellulitis

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Abstract ● Background: Lethal midline granuloma usually presents with rhinorrhoea and redness of the skin above the nose. Early ocular symptoms are very rare. We here describe a patient who presented with acute orbital cellulitis. ● Patient: A 73-year-old woman had a 24-h history of severe pain around her left eye. We saw the typical clinical picture of orbital cellulitis. A CT scan revealed a diffuse infiltration of the left upper and lower lid, the anterior orbit and the ethmoidal sinuses. ● Result: On surgical exploration we found a granular, partly necrotic tumour. Histological examination revealed an angiocentric nasal T-cell lymphoma (midline granuloma). ● Conclusion: Midline granuloma should be included in the differential diagnosis of acute orbital cellulitis.

"Lethal midline granuloma" comprises tumours in the midface with poor prognosis, including Wegener's granulomatosis, idiopathic midline destructive disease, lymphomatoid granulomatosis and angiocentric lymphoma [2]. Many cases of lethal midline granuloma are now believed to represent nasal T-cell lymphomas [4, 7, 8, 9]. Ocular involvement has rarely been reported in the midline granuloma syndrome apart from Wegener's granulomatosis. Clinical signs usually begin in the midface with a blocked nose and/or skin lesions. We describe a patient presenting with sudden lid swelling and vision loss caused by a midline angiocentric T-cell tumour.

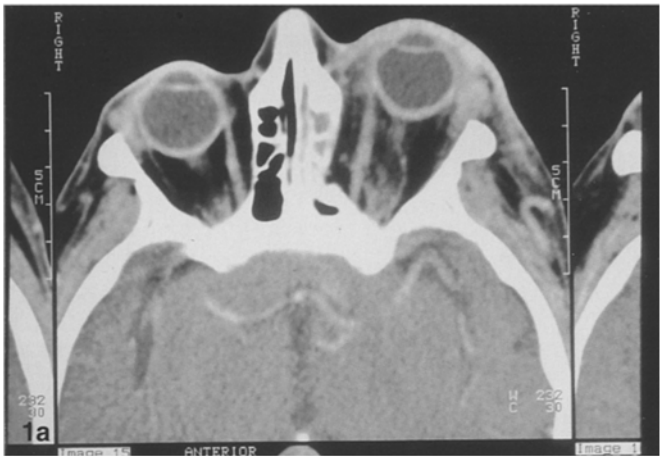
A 73-year-old white woman presented with a 24-h history of severe pain around her left eye. She was referred to our hospital as an emergency from a rehabilitation eye clinic, where she had been treated for chronic recurrent bilateral uveitis of unknown aetiology. The uveitis had first been diagnosed 13 years previously. Pars plana vitrectomy, cryocoagulation and laser coagulation of the peripheral fundus had been performed on the left eye at another eye clinic, as had bilateral cataract extraction.

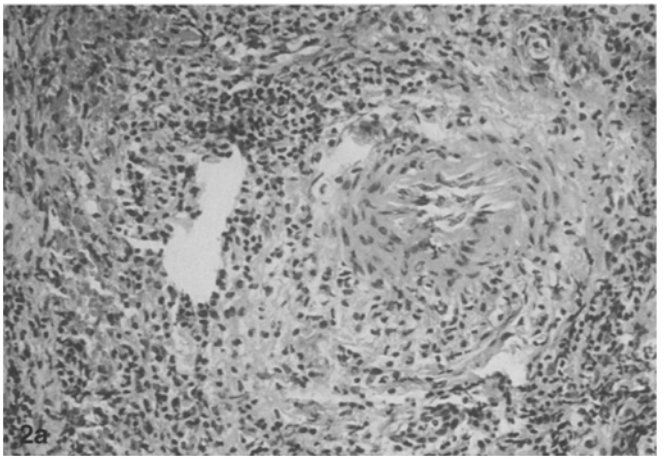
On her first visit she complained of having had a cold for 2 weeks and of headache in the area of the left eye. Generally she suffered from moderate arterial hypertension, gonarthrosis and depression.

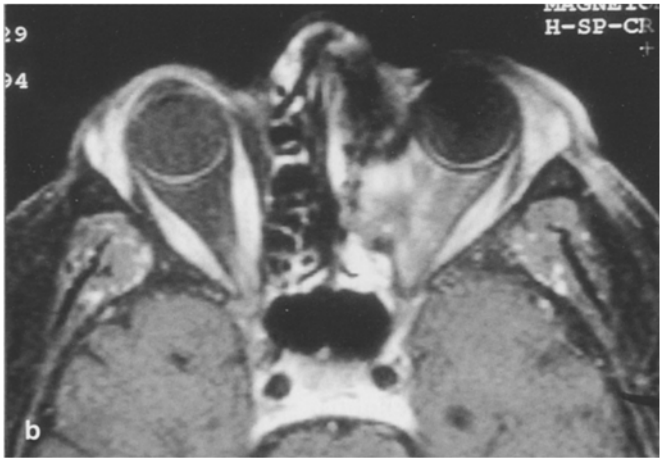
On examination we saw the typical clinical picture of left orbital cellulitis with massively swollen lids, marked chemosis and, as far as we could see, abolished ocular motility. The left globe could not be examined further; the right eye was normal. Body temperature was elevated (38.3° C). Blood cell count and routine laboratory parameters were normal except for slightly elevated bilirubin (1.15 mg/dl). Anticytoplasmic antibodies (ANCA) were negative, SD Wegener's granulomatosis was unlikely.

In the ENT department our diagnosis (orbital cellulitis) was confirmed. Endonasal hyperaemia and swelling of the mucosa were also found, as was moderate putrid rhinorrhoea. A CT scan performed the same evening showed diffuse infiltration of the left upper and lower lid, the anterior orbit and left ethmoidal sinuses (Fig. 1a). The medial orbital wall was partly destroyed, while the left globe itself appeared unaltered.

Systemic antibiotic therapy was started immediately, and surgical exploration of the left orbit and pansinusectomy were performed the same day. To our surprise we found dark, granular, partly necrotic tissue. The mucosae of the left nasal cavity and the left ethmoidal sinus were affected as well as the left orbit. Histological examination revealed a T-cell lymphoma with necrotic areas and infiltration of vascular walls ("angiocentric lym-







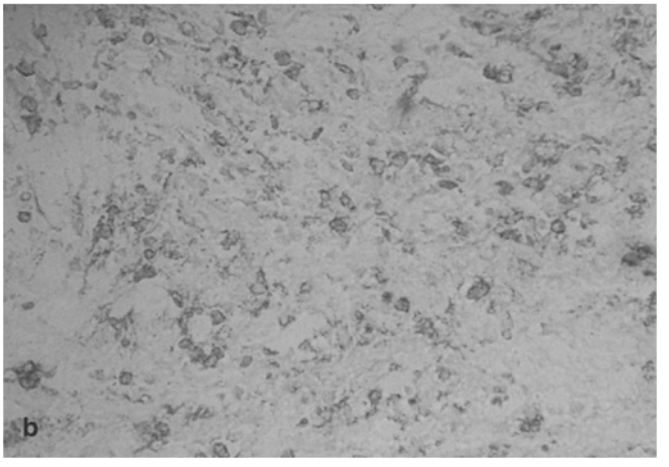


Fig. 1 **a** Transverse CT scan of the orbital region. Note infiltration in the left ethmoidal sinus and orbit. **b** MR image of the orbital region. Infiltration of the left ethmoid, orbit and lids. The left optic nerve is also infiltrated

Fig. 2 **a** A biopsy of the left orbit, showing a pleomorphic lymphoid infiltrate with invasion of the vascular sheaths, angiocentric t-cell lymphoma (hematoxylin and eosin, ×65). **b** Immunohistochemical staining of CD3-positive (T) cells within the removed tumour tissue (×130)

phoma", Fig. 2a). The cells were of middle size and showed marked polymorphism, immunohistochemical staining (3-amino-α-ethylcarbazole) showed CD3 positively (Fig. 2b).

On the first postoperative day, the left eye could also be examined because large parts of the tumour had been removed. We found a normal anterior chamber without cells and flare on both sides. Best corrected visual acuity was 5/10 (OD) and 2/10 (OS). Fundus examination revealed a bilateral macular pucker, sheathed venous vessels in the mid-periphery and, in the left eye, a swollen, pale optic disc with few parapapillary haemorrhages. The most likely clinical diagnosis was anterior ischaemic optic neuropathy (AION) due to infiltration of the optic nerve. The latter was affirmed by MR imaging (Fig. 1b). Fluorescein angiography revealed bilateral choroidal folds without venous leakage.

Radiotherapy (50 Gy) was performed over the next 5 weeks. The whole nasal cavity, both ethmoidal sinuses and the left orbit were within the radiation field. Under this therapy the lid swelling decreased and ocular motility partly returned. Disc swelling and choroidal folds decreased, but visual acuity remained unchanged. Apart from mild conjunctivitis of the left eye, ocular side effects of the radiation were not observed.

While in Wegener's granulomatosis ocular involvement is not uncommon and may be the first clinical sign [3], in the other subtypes of the midline granuloma syndrome patients typically present with upper respiratory tract problems. The first symptoms are usually mucous or serous rhinorrhoea, swelling and redness of the skin above the nose, followed by rapid and relentless destruction and deformation of the nose, the paranasal sinuses and the palate [5].

Secondarily, if at all, the patients may show affections of the eye or its adnexae. In one patient with idiopathic midline destructive disease, subcutaneous eyelid nodules were found [1], and in another patient dacryocystitis was reported [6]. In the latter case a poorly differentiated lymphoma of the midline granuloma type was the cause.

In these two cases the first clinical signs were seen in the throat and nose [6] and in outer parts of the nose [1].

Our patient presented with an angiocentric (T-cell) lymphoma of the midface. Histology and infiltration of the nasal cavity were typical for a "lethal midline granuloma". Like this case, most nasal lymphomas show CD3 positivity [4]. While it is not extremely uncommon for a large angiocentric lymphoma to finally invade the orbital wall [8], the presentation with orbital cellulitis is very unusual.

To the best of our knowledge this is the first published case primarily presenting with ocular signs. Our patient was admitted to hospital due to acute "orbital cellulitis". The clinical picture was dramatic and fully blown within 24 h. Infiltration of the paranasal sinuses was present at the same time. Unlike the orbital infiltration, however, these mucosal alterations remained almost asymptomatic. In our patient the medial bony orbital wall was obviously penetrated early in the disease, allowing the tumour to spread into the anterior orbit before symptoms due to tumour growth in the nasal cavity and the sinuses occurred. Presumably the tumour originated from the mucosa adjacent to the medial orbital wall.

Whether the uveitis was related to the midline granuloma remains unclear, but an association seems unlikely due to the long clinical course of the uveitis.

"Lethal midline granuloma" can present as an acute orbital inflammation and should be included in the differential diagnosis of acute orbital cellulitis.

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