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OCULAR DISEASE

Optic Nerve Glioma



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Introduction

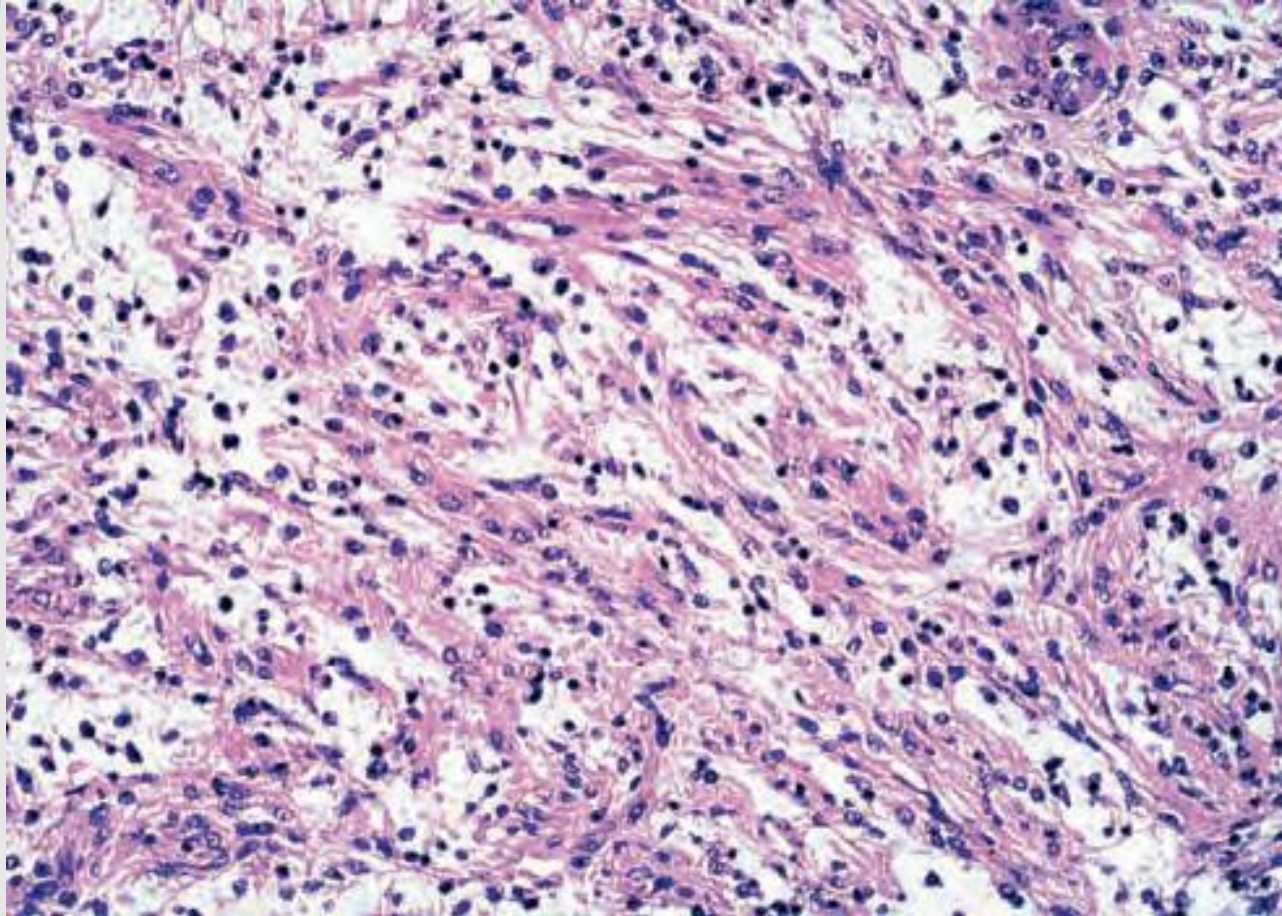
- Optic nerve glioma is a
 - ✓ slowly growing,
 - ✓ Pilocytic astrocytoma that typically affects children (median age 6.5 years);
 - ✓ Histopathology shows spindle-shaped pilocytic (hair-like) astrocytes and glial filaments



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Histopathology – spindle shaped pilocytic astrocytes and glial filaments



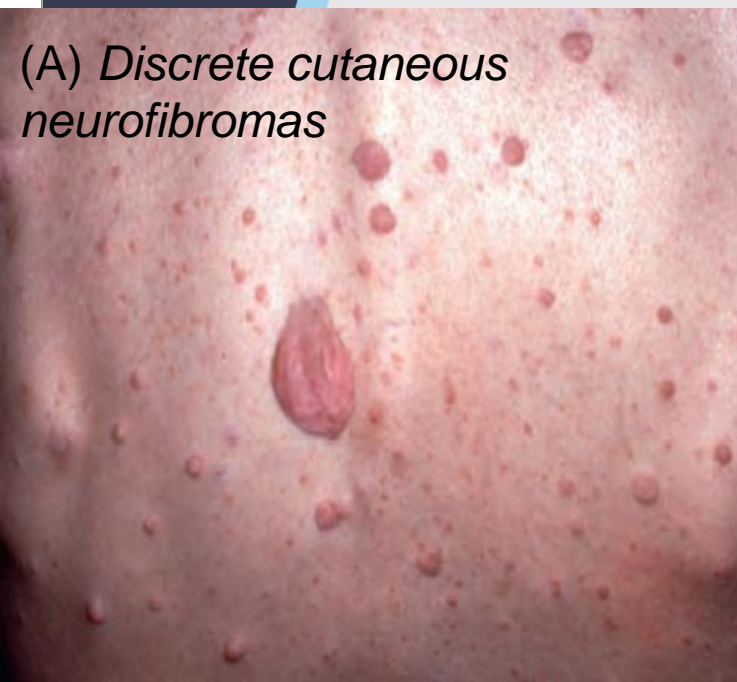


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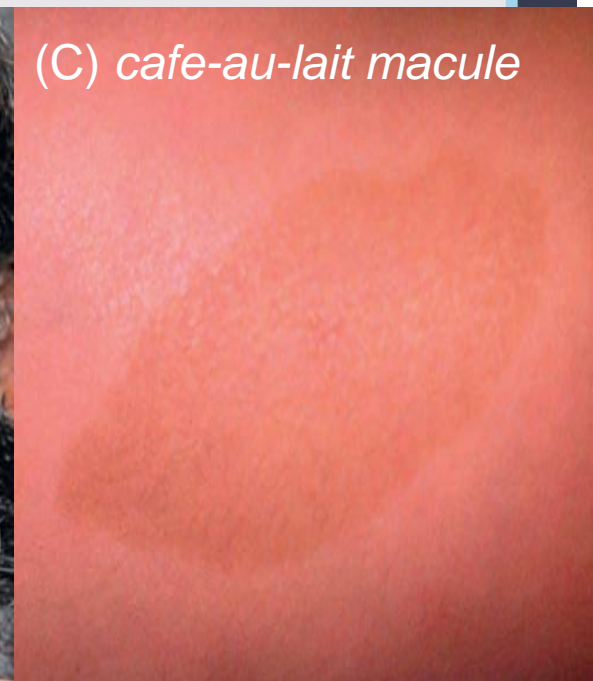
- Approximately 30% of patients have associated **Neurofibromatosis type I** and in these patients the prognosis is generally superior.



(A) *Discrete cutaneous neurofibromas*



(B) *Elephantiasis nervosa*



(C) *cafe-au-lait macule*



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- Malignant glioma (glioblastoma) is rare, has a very poor prognosis, and usually occurs in adult males.



Diagnosis

Symptoms

- Slowly progressive visual loss
- Proptosis
- Acute loss of vision due to haemorrhage into the tumour can occur

Signs

- Proptosis is often non-axial, with temporal or inferior dystopia
- Optic atrophic
- Opticociliary collaterals and central retinal vein occlusion
- Intracranial spread to the chiasm and hypothalamus may develop

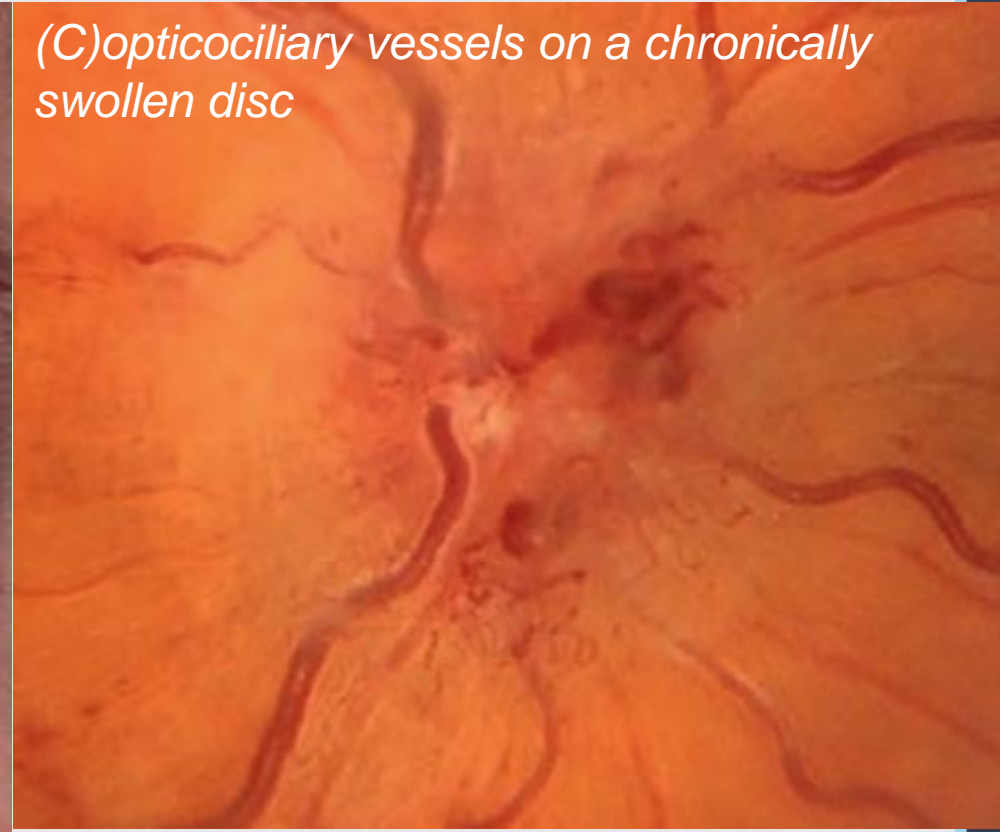


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(A) proptosis with inferior dystopia



(C) opticociliary vessels on a chronically swollen disc





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Investigation

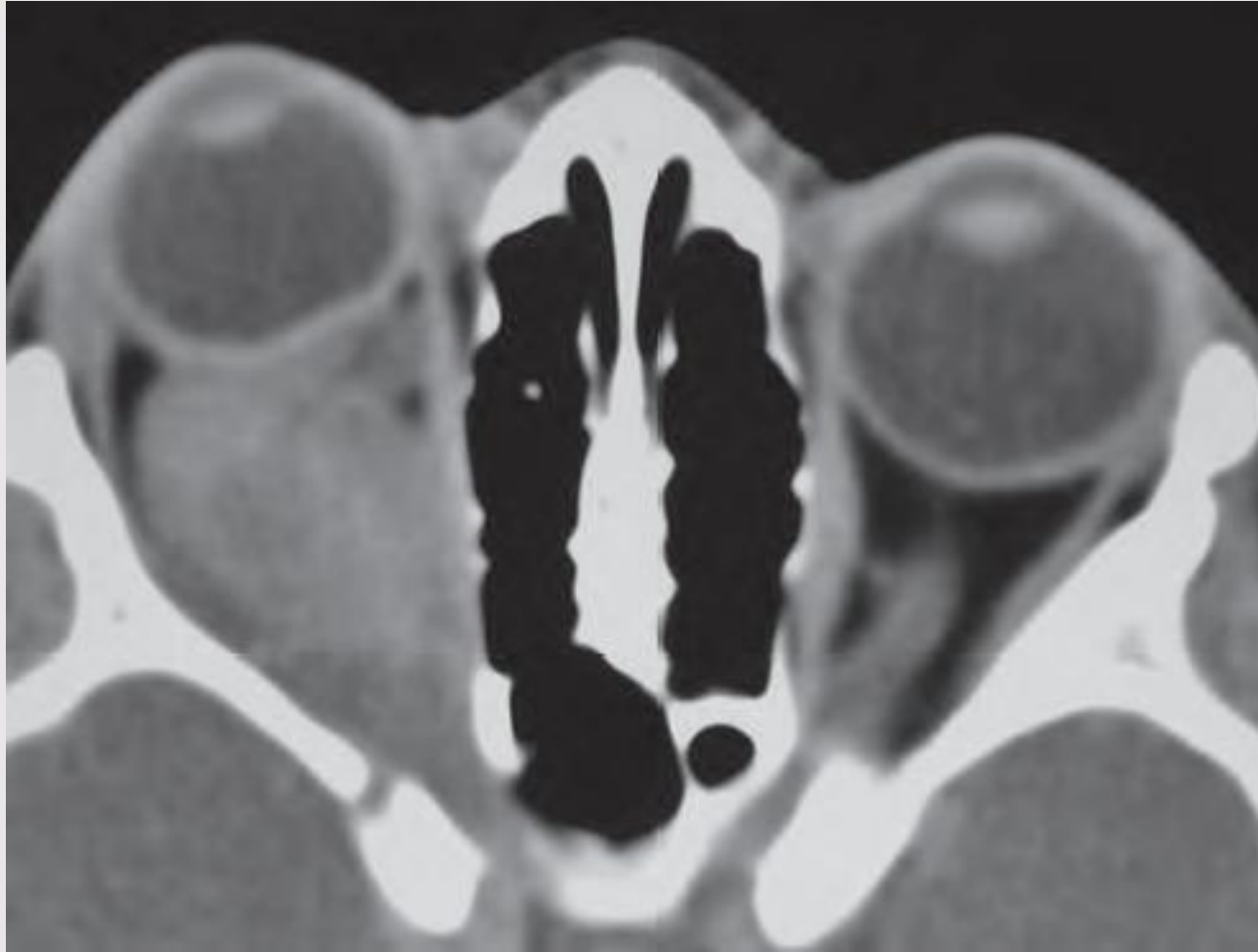
- MRI effectively demonstrates the tumour, and may show intracranial extension if present.
- CT in patients with associated NF1 shows a fusiform enlargement of the optic nerve with a clear-cut margin produced by the intact dural sheath.
- In patients without NF1 the nerve is more irregular and shows low-density areas



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Axial CT showing fusiform optic nerve enlargement





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Treatment

- ✓ **Observation** may be considered in patients with a typical pilocytic astrocytoma on imaging in whom the tumour is confined to the orbit, especially if there is good vision and no significant cosmetic impairment; serial MRI is important if this option is chosen. Spontaneous regression has been reported, usually in NF1, but is very rare



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- **Surgical excision** with preservation of the globe is indicated in those with large or growing tumours where complete resection of the tumour can be achieved, particularly if vision is poor and proptosis significant.



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- **Radiotherapy** may be combined with chemotherapy for tumours with complete surgical excision.