

Image Quiz

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Painless Grey Eyes

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A 68-year female, with rheumatoid arthritis for more than 20 years, presented with recent onset, progressive discolouration of both sclera. It was painless and she had no significant visual loss.



1. Identify the clinical signs demonstrated.

DIAGNOSIS

This is scleromalacia perforans or anterior necrotizing scleritis without inflammation [1,2]. The characteristic features are,

- Thinning, necrosis, and perforation of sclera
- Underlying dark uveal tissue becomes visible as grey patches.
- Dilated scleral vessels.

It is usually bilateral and asymptomatic and is an incidental clinical diagnosis during ophthalmological examinations.

DISCUSSION

Predominantly seen with long-standing rheumatoid arthritis, specially in elderly females. However, following multiple diseases are known to be associated with this condition [1].

- SLE
- Behcet disease
- granulomatosis with polyangiitis
- Polyarteritis nodosa
- Crohn disease
- Graft vs Host disease
- Porphyria
- Varicella zoster

Complications

- Globe rupture with minimal or no trauma
- Raised intra-ocular pressure.

Treatment

No effective treatment is available up to now. Considering the presumed vasculitic/autoimmune process, steroids, together with immunosuppressive medications [1,2] such as;

- Cyclophosphamide
- Methotrexate
- MMF
- Azathioprine is used.

Further, the newer immunomodulating medications, the biologics have been used with promising results [1].

- tumor necrosis factor inhibitors—TNF1 - etanercept, infliximab
- interleukin-1 receptor antagonist - anakinra
- interleukin-2 receptor blocker - daclizumab
- anti-lymphocyte medicament - rituximab, alemtuzumab

Surgical intervention with tectonic patch grafting is undertaken to maintain globe integrity.

REFERENCES

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