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Sir,

Orbital cellulitis in a patient with orbital neuroblastoma and febrile neutropenia

Neuroblastoma is a rare malignant tumour of neural crest cells, which can metastasize to the orbit and present as proptosis and periorbital ecchymosis ('raccoon eyes'). This is deemed stage 4 disease.[1] It is associated with an unfavourable prognosis, and management of this stage is with chemotherapy (followed by local excision and radiotherapy of the primary). Chemotherapy constitutes an induction phase followed by myeloablative conditioning with haematopoietic stem cell reconstitution and treatment for minimal residual disease.[2] The diagnosis of orbital cellulitis in these patients may be difficult to establish, in the setting of pre-existing proptosis from the metastasis and febrile neutropenia from immunosuppression. Such a case is presented here.

Case report

A 4-year-old girl was referred with headaches, right subconjunctival haemorrhage, and proptosis. An immediate CT scan revealed a right superolateral orbital mass (Figure 1a). A biopsy of the orbital mass by an ophthalmic surgeon confirmed neuroblastoma. The primary lesion was localized to the left adrenal gland with abdominal ultrasound and CT. Eighty days of the neuroblastoma high-risk 1 protocol comprising IV vincristine, carboplatin, and etoposide was commenced. During chemotherapy, recurrent episodes of febrile neutropenia developed. During an episode of febrile neutropenia she was noted to have deteriorating proptosis and development of periorbital erythema on day 70 of chemotherapy. The doubt was whether this represented treatment failure with deterioration of the orbital tumour or an orbital infection. A CT scan revealed a right pansinusitis, and right medial rectus enlargement indicating extension into the extraconal compartment. The original orbital mass showed no radiological deterioration and orbital cellulitis was deemed by the probable pathology. She was placed on intravenous teicoplanin, meropenem, and amphoterocin. A sinus washout was performed, which revealed a fungus. Thoracic CT demonstrated multiple areas of lung consolidation. Lung biopsy confirmed the organism to be *Aspergillus flavus*. Voricanozole was added to maximize antifungal therapy. Intravenous metronidazole and ciprofloxacin were added for concurrent urinary infection. A week later the preseptal inflammation improved but visual acuity decreased to light perception only and an RAPD developed. An urgent CT showed the right medial rectus compressing the optic nerve (Figure 1b). Urgent antral washout with ethmoidectomy and medial wall decompression was performed (Figure 1c). Within days there was marked systemic improvement, reduction of the proptosis and preseptal inflammation. The visual acuity stabilized at 2/60 with a residual pale optic disc secondary to optic nerve compression.

Comment

The clinical dilemma here was to ascertain treatment failure or orbital cellulitis, as the symptoms and signs of orbital cellulitis in this situation became of questionable value. The normal signs of orbital cellulitis are fever, proptosis, and periorbital erythema. All of these signs could be attributed to the primary disease or to the complications of treatment. A CT scan was invaluable as it demonstrated pansinusitis and extraconal orbital involvement with no unfavourable change in characteristics of the original tumour.

Between 10 and 20% of cases[3] of neuroblastoma have orbital involvement all of whom undergo chemotherapy. As orbital neuroblastoma and orbital cellulitis have very similar features during chemotherapy, a high index of

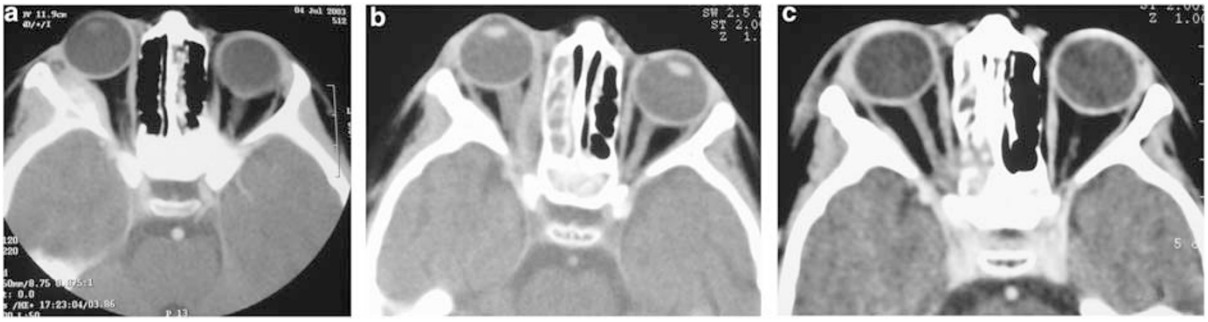

Figure 1 (a) the right lateral orbital tumour; (b) the right medial rectus compressing the optic nerve; (c) the appearance following orbital decompression and completion of chemotherapy.

suspicion is imperative in order not to miss a diagnosis of orbital cellulitis in this situation. Russo *et al*[4] have reported a case of orbital cellulitis in a patient with neuroblastoma without orbital metastasis. To the best of our knowledge, no presentation of orbital cellulitis in a case of metastatic orbital neuroblastoma has been hitherto reported.

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References

1 Brodeur GM, Pritchard J, Berthold F, Carlsen NL, Castel V, Castelberry RP *et al*. Revisions of the international criteria for neuroblastoma diagnosis, staging, and response to treatment [see comments]. *J Clin Oncol* 1993; **11**: 1466–1477.
2 Matthay KK, Villablanca JG, Seeger RC, Stram DO, Harris RE, Ramsay NK *et al*. Treatment of high-risk neuroblastoma with intensive chemotherapy, radiotherapy, autologous bone marrow transplantation, and 13-*cis*-retinoic acid. Children's Cancer Group. *N Engl J Med* 1999; **341**: 1165–1173.
3 Belgaumi AF, Kauffman WM, Jenkins JJ, Cordoba J, Bowman LC, Santana VM *et al*. Blindness in children with neuroblastoma. *Cancer* 1997; **80**(10): 1997–2004.
4 Russo G, Di Pietro M, La Spina M. Ocular involvement in neuroblastoma: not always metastasis. *Lancet Oncol* 2004; **5**(5): 324.

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Sir,

Recurrent subconjunctival and periorbital haemorrhage as the first presentation of systemic AL amyloidosis secondary to myeloma

Amyloidosis is the deposition of amyloid protein in organs and tissues. The occurrence of amyloid in the eye adnexal structures is well documented but uncommon. Systemic AL amyloidosis is a rare condition in which the monotypic amyloidogenic immunoglobulin light-chain molecule is thought to be the precursor protein. This may be secreted by monoclonal plasma cells infiltrating the tissue and is associated with monoclonal plasma cell proliferations such as myeloma. Clinically conjunctival amyloidosis usually presents as a painless palpable mass[1] and occurs most commonly after inflammatory conditions such as trachoma,[2] or more rarely with chronic recurring uveitis,[3] rheumatoid arthritis,[4] and Churg–Strauss syndrome.[5] Systemic forms of amyloidosis usually spare the conjunctiva.[6] Even with monoclonal gammopathies and multiple myeloma where systemic amyloidosis is common, conjunctival amyloidosis is rare.[7]

Case report

A 53-year-old man presented to an ophthalmology clinic with a history of recurrent sub-conjunctival haemorrhages and periorbital bruising occurring spontaneously or after minimal straining. He also complained of more recent constant diplopia. He had