

RETINOBLASTOMA WITH SKIN METASTASIS

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A rare case of bilateral retinoblastoma with cutaneous metastases in a 10-month-old male child with a rapidly fulminant course is being reported.

Key Words : Retinoblastoma, Skin metastasis

Introduction

Retinoblastoma is a malignant intraocular tumour of childhood which occurs in about 1 in 15,000 live births. It occurs in non familial forms, the latter being genetically transmitted by an autosomal dominant mode of inheritance. It is bilateral in 1/3 of the cases. The etiology is not entirely understood, but certain environmental factors as well as genetic factors appear to play a role in the development of the tumour.¹

Case Report

A 10-month-old male child presented with gradual protrusion of the right eye of 3 months duration; and numerous, erythematous and skin-coloured, cutaneous and subcutaneous, mobile, non tender nodules over face, scalp, upper limbs and upper trunk (Fig.1) of 2 months duration.

Initial white pupillary reflex (leucocoria) was noticed in the right eye 4 months back, which was

progressive. On ophthalmological examination a big fungating mass was seen at the site of the right eyeball involving both the upper and lower lids, and different parts of the eyeball could not be identified separately. Left eye revealed mild protrusion with anterior segment inflammatory signs. There was no evidence of strabismus or heterochromia of iris. Right eyeball was enucleated, and histopathology revealed undifferentiated retinoblasts with necrosis and calcification.

No similar history of white pupillary reflex was found among the other siblings. On examination of eyes of parents, the fundus showed no scar suggestive of retinoblastoma in their childhood. Routine hemogram, urinalysis, X-rays of skull and long bones, CSF examination and peripheral blood smear were within normal limits. Skin biopsy of the patient was refused by the parents, in view of the deteriorating health.

Discussion

Metastasis to skin is not common and when seen, implies a poor prognosis. Most metastases are seen on the head and neck, anterior chest wall and abdomen.²

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Retinoblastoma is diagnosed most frequently in children between 2 and 6 years of age. Approximately 10% of patients are diagnosed at birth. It most frequently metastasizes



Fig.1. Protrusion of right eyeball with cutaneous nodules over face, scalp, upper limbs and upper trunk.

to subcutaneous tissues of the head and preauricular lymph nodes. The most common cause of death is brain metastasis.³ Patients with retinoblastoma and their relatives appear to have an increased risk of other cancers, especially melanoma, which represents 7% of secondaries in retinoblastoma survivors.⁴

Direct involvement of the skin by metastatic spread from a distant primary tumour is unquestionably a marker for internal malignancy. This terminal case of retinoblastoma is probably the first case with cutaneous metastasis reported from India. The salient features were negative family history, bilateral affection, early age of presentation with fulminant course.

References

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