Case Report

Orbital rhabdomyosarcoma in an adult

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Abstract

Introduction: Rhabdomyosarcoma is the most common primary orbital malignant tumor in children. Orbital lesions represent about 10 % of all the cases of rhabdomyosarcoma. Rhabdomyosarcoma is a rare cause of proptosis in adults.

Objective: To report a case of primary orbital rhabdomyosarcoma in a 45-year-old female.

Design: Interventional case report. The main outcome measures are a rare cause of proptosis in an adult, discussion on treatment options and prognosis of rhabdomyosarcoma.

Result: The patient underwent total orbital exenteration and was referred for radiotherapy and chemotherapy.

Conclusion: Rhabdomyosarcoma is a rare cause of proptosis in adults. It should be suspected in a case of rapidly-progressive proptosis in adults.

Keywords: rhabdomyosarcoma, proptosis, malignant tumors, radiotherapy, chemotherapy, orbital exenteration

Introduction

Rhabdomyosarcoma (RMS) (from Greek, *rhabdo,* meaning rod shape, and *myo,* meaning muscle) is the most common soft tissue sarcoma in children. Though Weber first described RMS in 1854, a clear histologic definition was not available until 1946, when Stout recognized the distinct morphology of rhabdomyoblasts. Stout described rhabdomyoblasts as appearing in round, strap, racquet, and spider forms. As its name suggests, the tumor is believed to arise from primitive muscle cells (Cripe TP, 2008). Orbital Rhabdomyosarcoma was first reported by Bayer (Henderson JW, 1973).

Rhabdomyosarcoma is the most common childhood primary soft tissue sarcomas of the orbits (Porterfield JF & Zimmerman LE, 1962). Orbital lesions represent about 10 % of all the cases of rhabdomyosarcoma (Crist W et al, 1995). Rhabdomyosarcoma is a rare cause of proptosis in adults. We report a rare cause of proptosis due to rhabdomyosarcoma in a 45-year-old female.

Case report

A 45-year-old female presented with a gradual protrusion of the right eye of three-year duration. It was associated with gradually progressive diminution of vision for the last one year. The protrusion of the right eye was rapidly progressing for the last fifteen days, which was associated with pain, redness, discoloration of the central black part of the eye. There

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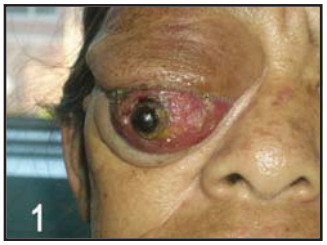

Figure 1
Right non-axial proptosis with exposure keratopathy and iris prolapse

was no history of trauma, fever or any systemic problems. She had no history of thyroid disorder. She was not commenced on any treatment prior to hospitalization.

Figure 1
Systemic examination did not reveal any abnormality. The best-corrected visual acuity was no perception of light (NPL) in the right eye and 6\6 in the left.

There was a non-axial proptosis (27mm) on the right side with the globe displaced infero-nasally. Palpation revealed a firm non-tender mass in the superior and lateral quadrants of the orbit. There was complete exposure of the cornea with restriction of extraocular movements in all directions of gaze. The conjunctiva was chemosed and injected. The cornea was melted down due to exposure keratopathy with prolapsed iris tissue at the centre, which was covered with an exudative membrane. There was complete destruction of the anterior segment of the right eye. The inner details were not visible. The left eye examination was within normal limits. The regional lymph nodes were not palpable.

The CT scan of the right orbit showed a homogenous orbital mass with erosion of the superior and lateral orbital walls and a deformed right globe with retinal detachment. The optic nerve sheath complex was normal.

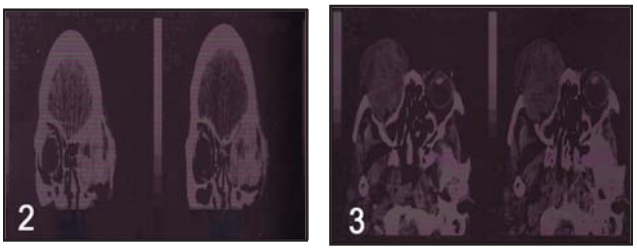

Figure 2 & 3
CT scan of orbit showing homogenous orbital mass with erosion of superior and lateral orbital walls; and a deformed right globe with retinal detachment. Optic nerve sheath complex was normal.

(Figure 2 & 3)
The patient underwent a right orbital exenteration under general anesthesia. Intra-operatively, involvement of superonasal part of the orbital roof with CSF leakage was detected. Intra-operatively, the patient was commenced on intravenous antibiotics. Histopathological analysis of the mass was conclusive of alveolar rhabdomyosarcoma. The patient was referred for radiotherapy and chemotherapy.

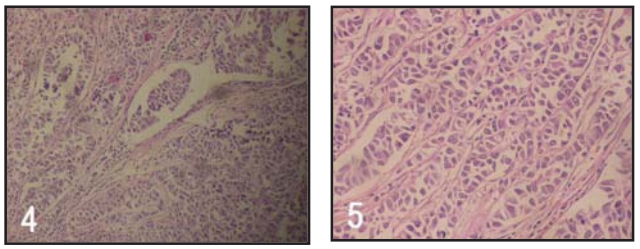

Figure 4 and 5
These figures show malignant round cell tumor revealing alveolar glandular trabecular and solid pattern with areas of necrosis and hemorrhages. The cytoplasm is deeply esinophilic with eccentrically placed nucleus.

Discussion
Rhabdomyosarcoma is a malignant neoplasm that is composed of cells with histologic features of striated muscle in various stages of embryogenesis (Weiss SW & Goldblum JR, 2001). It can occur in several sites in the body including the ocular region. Orbital rhabdomyosarcoma accounts for about 25-35 % of head and neck rhabdomyosarcoma and for about 10-20 % of all the cases of rhabdomyosarcoma (Crist W et al, 1995). Metastatic disease to the orbit from more distant body sites has been reported (Fekhat S et al 1993). Primarily, rhabdomyosarcoma is a disease of young children with a mean age of 8 years at diagnosis (Ashton N & Morgan G, 1965). However, it can occur at any age, with cases reported in newborns (Chams H et al 1987) and in infants and adults. It has been diagnosed in 35, 65 and 78-year-old patients (Sood GC et al 1970). It has also been diagnosed during pregnancy in two Nigerian women (Olurin O, 1969). Our patient presented at the age of the fifth decade. There is a slight predilection for disease in males, with a male-to-female ratio of 5:3 (Jones IS et al 1966).

Microscopically, the four major histopathological types of rhabdomyosarcoma are pleomorphic, embryonic, alveolar and botryoid. The majority of orbital rhabdomyosarcomas are of the embryonal type. The alveolar and botryoid types are less common, and the pleomorphic type is extremely rare in the orbit. Embryonal rhabdomyosarcoma is characterized histopathologically by spindle to round cells that show features of skeletal muscle in various stages of embryogenesis (Weiss SW, Goldblum JR, 2001). Our

case showed malignant round cells with closet morphologic resemblance to the alveolar type. Rhabdomyosarcoma shows strong tendency for local invasion, local recurrences and haematogenous and lymphatic spread. Therefore, treatment of Rhabdomyosarcoma includes a combination of both chemotherapy and radiotherapy (Demirci H et al 2002). According to the Intergroup Rhabdomyosarcoma Study Groups I through IV, prognosis of the disease depends on several factors like the extent of the disease, tumor burden at diagnosis, primary site, patient age, histopathologic and cytopathologic type, cellular DNA content and therapeutic response (Maurer HM et al 1988). With regard to tumor burden at diagnosis, survival rates are 90 % for patients with clinical group I disease, 85 % for group II, 70 % for group III and 40 % for group IV disease (Maurer HM et al 1988).Tumor morphologic features are an important predictor of death. Those patients with the alveolar cell type showed a 74 % 5-year survival, whereas those with the embryonal cell type demonstrated a 94 % 5-year survival (Lanzkowsky P, 2000). A younger age (1-7 years) at presentation carries better prognosis (Maurer HM et al 1988) than older age (>7 years). Infants particularly those of less than 1 year of age with orbital rhabdomyosarcoma show poor prognosis, with death in 46 %.The reason for more aggressive behavior of rhabdomyosarcoma in infancy is unknown (Kodet R et al 1997).

Until the late 1960s, orbital exenteration was generally considered to be the treatment of choice for orbital rhabdomyosarcoma (Jones IS et al 1966). However, the mortality rate for patients with orbital rhabdomyosarcoma continued to be greater than 70 % (Knowles DM et al 1976). Hence, orbital exenteration alone is rarely performed as a primary treatment today. It may be justified for extremely advanced disease that has destroyed the eye, as frequently seen in third-world and tumors resistant to irradiation and chemotherapy (Shields CL et al 2001). The Intergroup Rhabdomyosarcoma Study Committee (IRSG, the later Group) organized in 1972, performed large collaborative randomized trials for treatment of rhabdomyosarcoma. Since its inception, there have been four major trials, listed as studies I through IV (Crist W et al1995). As a result of these trials, survival after treatment of rhabdomyosarcoma at all sites has improved from 25 % in 1970 to 70 % in 1991 (Crist WM, Kun LE, 1991). Orbital rhabdomyosarcoma has been recognized to display better life prognosis than rhabdomyosarcoma at other sites. Before the treatment trials, patient survival with orbital rhabdomyosarcoma was poor, with approximately 30 % survival (Knowles DM et al 1976). Treatment generally consisted of orbital exenteration and various chemotherapy regimens. After trials I and II, improved treatment regimens with chemotherapy and radiotherapy, usually avoiding exenteration, were successful, and the prognosis of orbital rhabdomyosarcoma strikingly improved to 93 % survival at 3 years (Wharam M et al 1987).

According to the results of trial IV, the recommended treatment includes both chemotherapy and radiotherapy, with the exception of completely resected orbital tumors where only chemotherapy without radiotherapy is advised. Treatment depends upon the group of the disease at presentation as classified by the IRSG. Current management of group IV orbital rhabdomyosarcoma depends on the location and extent of disease and generally consists of a combination of chemotherapy and radiotherapy delivered to the orbit and all involved sites of the tumor (Lanzkowsky P, 2000). Recurrent tumors in the orbit are usually treated with orbital exenteration, sometimes supplemented with chemotherapy and radiotherapy (Mannor GE et al 1997).

The prognosis for patients with orbital rhabdomyosarcoma has greatly improved in recent years. Based on trials I, II, III, and IV, survival with orbital rhabdomyosarcoma is now 93 % (Kodet R et al 1997).

Our patient at the time of presentation had an extensive tumor with destruction of the globe and erosion of the superior and lateral walls. Therefore she underwent orbital exenteration. Despite the rarity of this tumor at this age, the possibility of occurrence of rhabdomyosarcoma in elderly people cannot be ignored as is evident from our patient. Our clinical findings confirmed the diagnosis of rhabdomyosarcoma, and the patient was treated surgically and referred for radiotherapy and chemotherapy. The treatment outcome is not known to us because the patient was lost for follow-up and did not respond to written communication to her permanent address.

Conclusion

Rhabdomyosarcoma is a rare cause of proptosis in adults. However, it should be suspected as a cause of rapidly progressing proptosis in adults.

References

Ashton N, Morgan G (1965). Embryonal sarcoma and embryonal rhabdomyosarcoma of the orbit. T Clin Pth 1965;18:699-714

Chams H, Rahisii F, Sagdeghi- Tari A (1987): A case of congenital rhabdomyosarcoma. Orbit;7:133-5

Crist WM, Kun LE (1991). Common solid tumors of childhood. N Engl J Med;324:461-71.

Crist W, Gehan EA, Ragab AH et al (1995). The third inter-group rhabdomyosarcoma study. J Clin Oncol; 13: 610-630.

Demirci H, Shields CL, Shields JA (2002). Orbital tumors in the older adult population. Ophthalmology; 109:243–8.

Fekhat S, Miller NR, Lovry MC: Alveolar rhabdomyosarcoma that metastasized to orbit. Arch Ophthalmol III: 1993;1662-1664

Henderson JW (1973). Orbital tumors. 1st sub-edition. W.B. Saunders, Philadelphia: 269-84.

Jones IS, Reese AB, Kraut J (1996). Orbital rhabdomyosarcoma. Am J Ophthalmol; 61:721-36.

Kodet R, Newton WA Jr, Hamoudi AB et al (1997). Orbital rhabdomyosarcomas and related tumors in childhood: relationship of morphology to prognosis—an intergroup Rhabdomyosarcoma Study. Med Pediatr Oncol; 29: 51–60.

Knowles DM, Jakobiec FA, Potter GD, Jones IS (1976). Ophthalmic striated muscle neoplasms. Surv Ophthalmol; 21: 219-61.

Lanzkowsky P (2000). Manual of pediatric hematology and oncology, 3rd ed. New York: Academic Press; 527–53.

Mannor GE, Rose GE, Plowman PN et a (1997). Multi-disciplinary management of refractory orbital rhabdomyosarcoma. Ophthalmology 1997;104:1198-201.

Maurer HM, Beltangady M, Gehan EA et al (1988). The Intergroup Rhabdomyosarcoma Study - I. A final report. Cancer; 61: 209–20.

Olurin O (1969). Orbital rhabdomyosarcoma in pregnancy. Cancer; 24:1013-6.

Porterfield JF, Zimmerman LE (1962). Rhabdomyosarcoma of orbit: A clinicopathologic study of 55 cases. Virchows Arch [A]; 335:329-344.

Shields CL, Shields JA, Honavar SG, Demirci H (2001). Clinical spectrum of primary ophthalmic rhabdomyosarcoma. Ophthalmology;108:2284-92.

Sood GC, Sen DK, Diwan R, Aurora AL (1970). Orbital rhabdomyosarcoma in an infant. Br J Ophthalmol; 54:203–5 .

Cripe TP (2009). Rhabdomyosarcoma. eMedicine (accessed on 23.08.2009)

Weiss SW, Goldblum JR (2001). Rhabdomyosarcoma. In: Weiss Sw, Goldblum

JR.eds. Enzinger and Weiss's soft tissue tumors. St. Louis, CV Mosby Co. Edition;4: 785-835.

Wharam M, Beltangady M, Hays D et al (1987). Localized orbital rhabdomyosarcoma. An interim report of the Inter-group Rhabdomyosarcoma Study Committee. Ophthalmology; 94:251-4.

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