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CASE REPORT Open Access

Alveolar Rhabdomyosarcoma of the foot metastasizing to the Iris: report of a rare case

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Abstract

Background: Intraocular iris rhabdomyosarcoma is extremely rare, and in the 3 cases reported to date occurred as the primary site of tumour growth. We report a case of rhabdomyosarcoma of the foot metastasizing to the iris.

Case presentation: An 18-year-old white female was referred to the London Ocular Oncology Service for management of a metastatic rhabdomyosarcomatous deposit in the iris, a metastasis from alveolar rhabdomyosarcoma of the foot. She was diagnosed nearly 2 years earlier with the primary sarcoma with extensive systemic spread and treated by resection of the foot lesion and chemotherapy, and achieved a partial remission. The left iris deposit was noted while she was receiving systemic chemotherapy, heralding a relapse. However, anterior uveitis and raised intraocular pressure developed and she was referred to our service for further management. A left iris secondary rhabdomyosarcoma deposit was noticed and in addition a lacrimal gland mass, as indicated by ultrasound B scan of the eye and orbit. The patient was treated with external beam radiotherapy to the globe and orbit, but died 2 months after treatment completion.

Conclusion: Rhabdomyosarcoma of the iris is very rare and was previously documented only as a primary malignancy in this location. We report that secondary spread to the iris can also occur, in this case as the first sign of widely disseminated systemic relapse.

Keywords: Rhabdomyosarcoma, Iris, Metastasis, Case report

Background

Rhabdomyosarcoma (RMS) is the most common soft tissue sarcoma in the paediatric population [1]. The orbit is the primary tumour site in 10 % of cases and is rarely a site for secondary spread from a distant extra-orbital origin [2]. Intraocular primary RMS of the uvea is yet another rare presentation of the disease, described only in a handful of case reports [2]. Herein, we report a unique case of secondary RMS to the iris, a metastasis from alveolar RMS of the foot.

Case presentation

An 18-year-old white female was referred to the London Ocular Oncology Service for management of a metastatic RMS deposit of the left iris. She was originally diagnosed by the Oxford Oncology team with metastatic alveolar RMS 22 months earlier, with the primary in the right flexor digitorum brevis, with extensive metastatic disease, including intramuscular deposit to the calf, popliteal fossa nodes, external inguinal chain, multiple mediastinal nodes and bilateral lung parenchymal involvement with pleural effusion. Systemic chemotherapy (ifosfamide, vincristine, actinomycin and doxorubicin) with surgical resection of the right foot lesion, followed by maintenance chemotherapy achieved prolonged disease control.

Fourteen months after diagnosis of the metastatic RMS she noticed a change in the left iris colour. Examination done at the Oxford Eye Hospital indicated an iris mass (Fig. 1a), and on PET scan, extensive systemic relapse was detected, including uptake of the primary foot site, pulmonary nodules as well as the left iris. Vincristine, irinotecan and temozolomide (VIT) were started, to control the recurrent disease, including the iris metastasis, which showed

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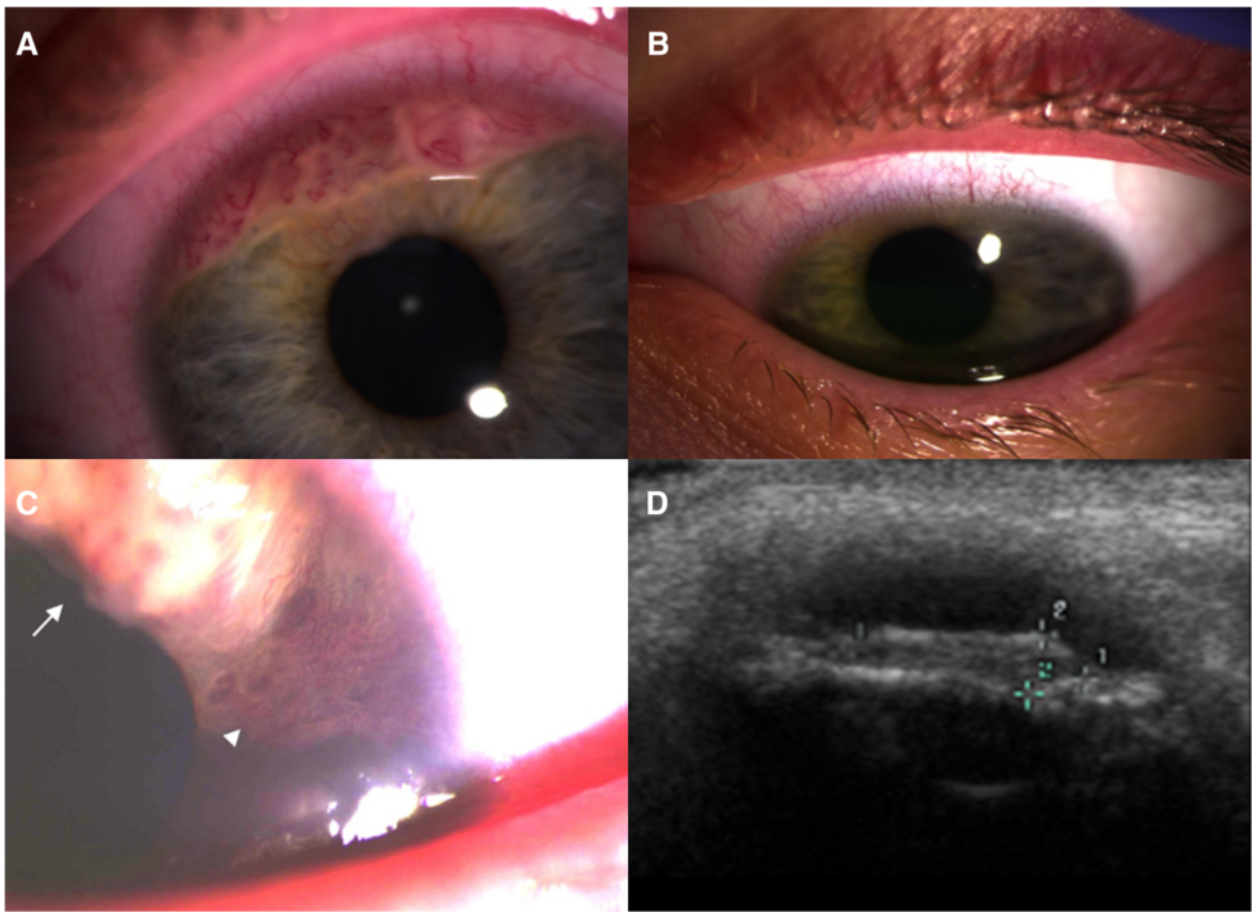

Fig. 1 Left eye iris mass at presentation (**a**), resolved after initial systemic chemotherapy (**b**). Ocular tumour relapse (*arrow*) and neovascularization of the iris (*arrowhead*) (**c**), demonstrated also on B mode ultrasound scan (**d**)

good response (Fig. 1b). A subsequent on – treatment relapse occurred after 6 cycles of VIT (primary site, lungs and musculoskeletal). Palliative oral etoposide was commenced 5 months after first intraocular involvement, keeping the disease under relative control.

After 3 months of palliative oral etoposide, the iris tumour recurred, with neovascularization (Fig. 1c), anterior uveitis and intraocular pressure of 40 mmHg. She was treated with topical antihypertensives and steroids and referred to our service for consideration for radiotherapy.

Visual acuity was 6/6 in the right eye and 6/36 in the left eye. Intraocular pressures were 14 mmHg RE and 15 mmHg LE. The right eye was entirely normal. The left eye had a ciliary flush. There were keratic precipitates but also larger areas of more confluent deposits on the corneal endothelium, with stromal oedema. An amelanotic multifocal vascularised mass was present in the superior and inferior aspect of the iris, occluding most of the iridocorneal angle. There were no ciliary body or choroidal tumours, but cupping of the optic disc was noted. B mode ultrasound scan (Acuson Sequoia 512, Siemens AG, Munich, Germany) with a 14 MHz linear B-scan array probe indicated a 1.7 mm elevated iris lesion with irregular anterior borders (Fig. 1d) and no ciliary body involvement, and in addition a lacrimal gland mass was noted. The patient was treated with palliative external beam radiotherapy (20Gy in 5 fractions) for both presumed intraocular and orbital metastatic RMS deposits. Treatment was well tolerated and there was a prompt and sustained clinical benefit. She died 2 months later (2 years after RMS diagnosis) from systemic progressive disease.

Discussion

There is a wide range of tumours that occur in the iris. Metastasis to the iris is infrequent, reported in fewer than 10 % of intraocular secondary deposits by Shields et al. and Konstantinidis et al. [3, 4]. In both reports the most common primary site was breast and lung, and there were no cases of systemic RMS metastasizing to the eye; hence the present case is highly unusual.

In another series by Shields and colleagues, the clinical features of 104 patients with iris metastasis from systemic

cancer were reported [5]. The median age was 60 years, most were white females, the main symptoms were pain or blurred vision and the main findings were corectopia and secondary glaucoma. Most tumours were unifocal and found in the inferior quadrant. In the present case, the patient was 18 years old when the intraocular mass was detected; she had no ocular symptoms but incidentally noticed iris heterochromia; the initial iris relapse was in the superior quadrant and only further relapse occurred superiorly and inferiorly, causing secondary glaucoma. In agreement with other reports, metastasis to the iris is a feature of advanced disseminated cancer with poor life prognosis [5].

RMS of the iris has been previously described, but is considered to be an extremely rare occurrence, reported to date only in 3 patients [6–8]. In all of these cases, the iris was the primary site of tumour growth. The Table summarizes the main clinical features found in those cases and in the present one. The age at presentation of patients with primary iris RMS was 5 years or younger and the presenting feature was development of an iris mass. Definitive treatment in those cases ultimately was enucleation with no local or systemic sequelae. The present case differed from the primary iris RMS ones in nearly all clinical features, but the mode of presentation. Management and outcome were obviously different.

RMS is classified into 4 histopathological types: embryonal, alveolar, botryoid and pleomorphic [2]. Orbital alveolar type is considered less common but more aggressive than the embryonal type [9]. The relation between the cell type, incidence and prognosis of intraocular RMS is not well established, since this is a rare occurrence.

Conclusion

Rhabdomyosarcoma of the iris is rare. Not only can this tumour develop in the iris as a primary site [10], but we report that secondary spread to the iris can also occur, in this case as the first sign of widely disseminated systemic relapse.

Abbreviations

RMS, rhabdomyosarcoma

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Availability of data and materials

All data presented in the manuscript.

Authors' contributions

IDF acquired the data, wrote the first draft of the manuscript and did a literature review, DH conceived of the study and edited the draft significantly, SW acquired the data and participated in the design of the study and its coordination, TF acquired the data and participated in the design of the study and its coordination, and MSS conceived of the study, participated in the design of the study and its coordination, did a literature review and revised the manuscript critically. All authors read and approved the final manuscript.

Competing interests

The authors declare that they have no competing interests.

Consent for publication

Consent to publish this case report was obtained from the patient's parents. A copy of the consent and all data and materials are available for review by the Editor-in-Chief of this journal.

Ethics approval and consent to participate

This report adhered to the tenets of the Declaration of Helsinki and was approved by the Moorfields Eye Hospital ethics committee. Consent to participate in this study was obtained from the patient's parents.

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