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BRIEF REPORT
Neonatal Prostatic Rhabdomyosarcoma

Amar Shah, MS, MCh, Karan Parashar, FRCS, Martin English, MD, and Harish Chandran, FRACS*

Rhabdomyosarcoma is one of the commonest soft tissue sarcomas of childhood, but neonatal presentation is extremely rare. This limited experience means there are no clear treatment guidelines in this age group. The authors report a boy with recurrent attacks of hematuria commencing in the neonatal period, which were shown to be from a prostatic rhabdomyosarcoma. To the best of our knowledge this is the first reported case of neonatal rhabdomyosarcoma in this site.Pediatr Blood Cancer 2004;43:275–277.

Key words: neonate; prostate; rhabdomyosarcoma

INTRODUCTION

Rhabdomyosarcoma is a tumour of mesenchymal origin, which can develop in any organ or tissue, other than in bone. It accounts for 4–8% of all malignant disease in children [1]. Approximately 5–10% of these are diagnosed during the first year of life [2]. Histology, group and tumour size may not be of such significant prognosis in the newborn as in older infants and children [3].

CASE REPORT

A 6-week-old male child was referred to us with a mass in the hypogastrium, which was first noticed by his father. The pregnancy had been uncomplicated, and there was no report of oligohydroamnios on antenatal ultrasound scans. The child was born by normal vaginal delivery at term. There was a history of two episodes of hematuria associated with difficulty in passing urine from the age of 3 weeks. On examination, he did not have any dysmorphic features. The bladder was distended up to the umbilicus. The rest of his abdomen was normal to examination. A 3×3 cm firm mass was felt below the bladder per rectum. Ultrasound examination showed a dilated thick-walled bladder with bilateral hydronephrosis. Urethral catheterisation was attempted unsuccessfully. Suprapubic catheterisation was, therefore, performed. An MRI scan of the pelvis was carried out which showed the presence of a well-defined mass at the base of the bladder surrounding the prostatic urethra measuring $1.9 \times 1.2 \times 3.9$ cm (Fig. 1). Cystoscopy confirmed the presence of a mass, which had invaded the prostatic urethra, with two polypoid growths projecting into the urethral lumen. The bladder was grossly trabeculated. Histopathological examination of the mass showed an alveolar rhabdomyosarcoma of the prostate (Fig. 2). One perivesical lymph node contained tumour. Bone marrow aspiration also showed invasion by tumour cells. Bone scan did not reveal any obvious skeletal metastases.

The patient was too young for treatment on the International Society of Paediatric Oncology (SIOP) Europe MMT 98 protocol for metastatic rhabdomyosarcoma. Treatment was based around the six-drug arm of the above protocol with the substitution of cyclophosphamide for ifosfamide because of the risk of nephrotoxicity, and the gradual introduction and dose escalation of actinomy-

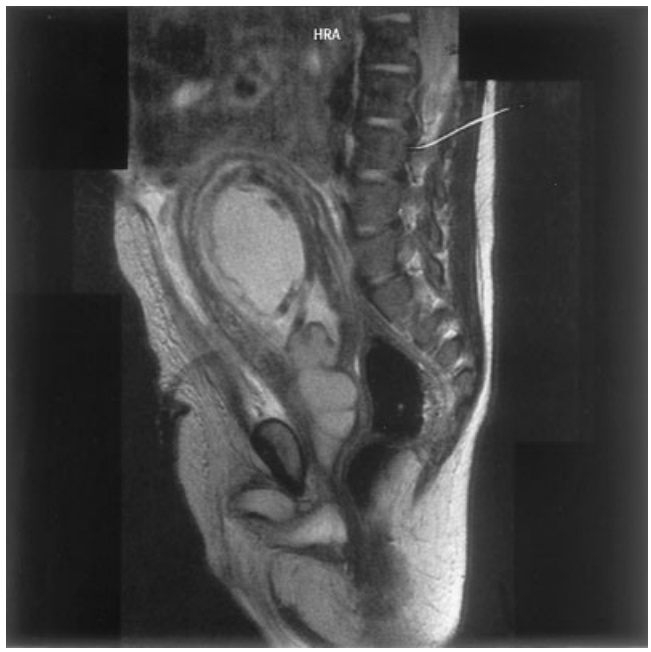

Fig. 1. Picture of the MRI showing the mass.

Birmingham Children's Hospital, Birmingham, United Kingdom

*Correspondence to: Harish Chandran, Consultant Paediatric Urologist, Birmingham Children's Hospital, Steelhouse Lane, Birmingham B4 6NH, United Kingdom. E-mail: Harish.Chandran@bch.nhs.uk

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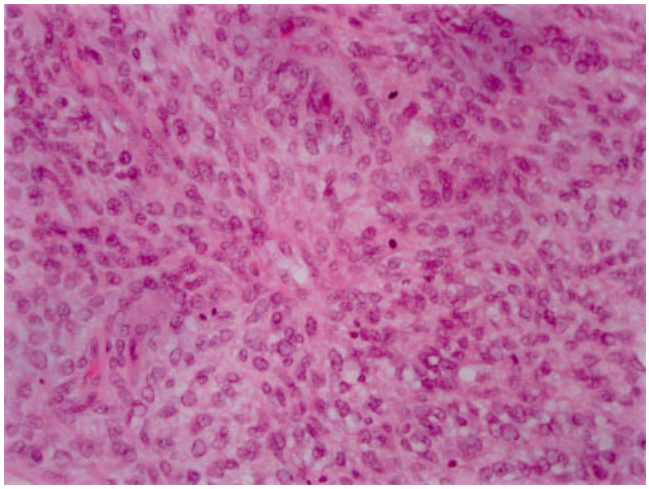

Fig. 2. Histology of the lesion suggestive of rhabdomyosarcoma at 200× magnification. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

cin, epirubicin, and etoposide in addition to vincristine and cyclophosphamide. Carboplatin was dosed based on renal function measured by ^{51}Cr-EDTA clearance to an area under the plasma concentration versus time curve (AUC) of 4.4 mg/ml min. This is equivalent to 250 mg/m^2. Details of the chemotherapy and toxicity with each course are shown in Table I.

He had a partial response to treatment with clearance of metastatic disease in bone marrow and lymph nodes and reduction in the size of the primary tumour to 1.7 × 1.4 × 1.4 cm. A total cystectomy was performed at 38 weeks of age with bilateral ureteroileostomy and ileal conduit. Histology confirmed treated alveolar rhabdomyosarcoma with clear histological margins. Maintenance chemotherapy with cyclophosphamide, vincristine, and actinomycin is continuing for a total of nine courses.

TABLE I. Chemotherapy and Associated Toxicity

		Toxicity grade (CTC)				
Course (week)	Drugs	Hb	Platelets	WCC	Infection	Other
1 (1–3)	VCR 0.025 mg/kg days 1, 8, 15 Cyclo 10 mg/kg days 1, 2, 3	2	0	0	0	0
2 (4–6)	VCR 0.025 mg/kg days 1, 8, 15 Cyclo 10 mg/kg days 1–5	2	0	2	0	SGOT grade 1
3 (7)	VCR 0.025 mg/kg day 1 Cyclo 10 mg/kg days 1–5 Act D 0.025 mg/kg day 1	2	0	1	0	0
4 (10)	VCR 0.033 mg/kg day 1 Carbo AUC 4.4 Epi 75 mg/m^2	2	2	3	0	0
5 (13)	VCR 0.033 mg/kg day 1 Cyclo 66 mg/kg day 1 VP16 75 mg/m^2 day 1	3	0	3	0	0
6 (16)	VCR 0.033 mg/kg day 1 Cyclo 66 mg/kg day 1 Act D 0.033 mg/kg day 1	2	1	3	0	0
7 (19)	VCR 0.05 mg/kg day 1 Carbo AUC 4.4 Epi 112.5 mg/m^2	2	0	1	0	0
8 (22)	VCR 0.05 mg/kg day 1 Cyclo 66 mg/kg day 1 VP16 112.5 mg/m^2 days 1 2	3	1	4	0	Constipation grade 2
9 (25)	VCR 0.05 mg/kg day 1 Cyclo 66 mg/kg day 1 Act D 0.033 mg/kg day 1	2	0	2	0	SGOT grade 1
10 (31)	VCR 0.05 mg/kg day 1 Cyclo 66 mg/kg day 1 Act D 0.045 mg/kg	2	2	4	0	0
11 (34)	VCR 0.05 mg/kg day 1 Cyclo 66 mg/kg day 1 VP16 112.5 mg/m^2 days 1–3	3	2	4	UTI + fever grade 2	0
12 (37)	VCR 0.05 mg/kg day 1 Carbo AUC 4.4 Epi 112.5 mg/m^2	2	0	0	0	0
M1 (40)	VCR 0.05 mg/kg Cyclo 750 mg/m^2 Act D 0.045 mg/kg	0	0	3	UTI grade 2	Constipation grade 2

CTC toxicity grades. Left ventricular fractional shortening noted to be 27% after course 11, but left ventricular function described as normal on observation.

DISCUSSION

The most common pelvic neoplasms in infants are germ cell tumours, especially sacrococcygeal teratomas, and rhabdomyosarcomas of the bladder. The Intergroup Rhabdomyosarcoma Study (IRS) Group was formed in 1972 to investigate the biology and treatment of rhabdomyosarcoma and undifferentiated sarcoma of soft tissue diagnosed during the first two decades of life. The favourable prognostic factors proposed by IRS in 1991 are [4]:

1) no distant metastases detectable at diagnosis,
2) primary sites in orbit and non parameningeal head/neck and genitourinary, excluding bladder/prostate regions,
3) grossly complete surgical tumour excision,
4) embryonal/botryoid histology,
5) tumour size <5 cm,
6) age younger than 10 years at diagnosis.

Our patient had a diagnosis of a stage 4 alveolar rhabdomyosarcoma of the prostate. The tumour was not resectable without major loss of organ function. Initial biopsy was followed by chemotherapy, with a plan for delayed surgery. Two percent of all childhood tumours occur in the first month of life and 20% of these are mesenchymal in origin [3]. Only 0.4% of the patients registered in the IRS studies I to IV were neonates.

Rhabdomyosarcoma presenting in the neonatal period have been reported in other sites, such as the tongue [5], neck [6], posterior trunk [7], thigh [8] as well as from the bladder and perineal region [9]. Only 5% of all childhood rhabdomyosarcomas arise in the prostate [10]. It commonly presents with obstructive urinary symptoms or an abdominal mass in an older child. A rhabdomyosarcoma protruding from the urethral meatus has been reported, in a 1-year-old [11].

There is an increased risk of actinomycin-D induced hepatic veno-occlusive disease in babies under 1 year and sudden cardiac death has been noted in a neonate treated with anthracyclines [12]. Most groups give guidelines for treatment using low-dose chemotherapy to attempt to reduce tumour size while the child grows older. Our patient had adverse disease stage and histology. Although the tumour was less than 5 cm in diameter, this was in a 5.6 kg, 7-week-old child, so it was proportionately a big tumour. In these circumstances, a more intensive approach was taken. Given that he is in complete remission post surgery, we hope that the chance of cure lies between 25 and 50%. We have elected not to give pelvic radiotherapy because of the late effects that this would cause in such a young child.

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