Rhabdomyosarcoma Arising in Teratoma of the Testis

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SUMMARY: Rhabdomyosarcoma arising in teratoma of the testis is an exceptionally rare occurrence. The rhabdomyosarcomatous element infers a less favourable prognosis and standard treatment regimes well recognised for the treatment of teratoma of the testis may not be effective.

Introduction

The development of non germ cell malignancy within gonadal or extragonadal teratoma is a rare occurrence. The majority of these tumours are sarcomas and rhabdomyosarcoma is seen predominately. Such tumours may respond poorly to standard Cisplatin based combination chemotherapy.

We report a case of rhabdomyosarcoma arising within a malignant teratoma intermediate of the testis and present a review of the literature.

Case Report

A 26-year-old officer presented with a 5 month history of gradual, non tender swelling of the right testis. He gave a history of two previous episodes of right sided epididymitis which had settled on antibiotics. There was no history of previous cryptorchidism. On examination the right testis was swollen to 15cm in diameter and was smooth, hard and non tender. The swelling failed to transilluminate and there was no associated palpable lymphadenopathy. Ultrasound examination revealed cystic changes in keeping with neoplasm and he went on to right radical orchidectomy. The pre-operative AFP was raised at 14 kU/L (Ref. range: 0.8 to 6.5 kU/L) but the Beta HCG was within normal limits. The histology demonstrated a mixture of undifferentiated teratomatous and differentiated teratomatous elements with large areas of rhabdomyosarcomatous stroma.

Staging investigations including Chest X-ray and CT scan of the chest and abdomen were unremarkable confirming the presence of stage 1 disease. After consideration it was decided not to offer adjuvant therapy but to maintain surveillance with monthly follow up and tumour markers together with 3 monthly CT scans. At follow up at 8 months there is no sign of recurrence.

Discussion

Rhabdomyosarcoma is an extremely rare non germ cell malignancy of the testis which may occur in combination with testicular germ cell tumour (1) or as pure rhabdomyosarcoma (2).

Accurate histological diagnosis is important and can be complicated in some instances as the features of sarcoma may resemble those of undifferentiated teratoma. Definitive diagnosis in doubtful cases may be made with immuno-histochemical studies which will be positive for myoglobin in the presence of rhabdomyosarcoma.

The origin of non germ cell malignancies within germ cell tumours is uncertain but may occur by malignant transformation of pre-existing teratomatous elements or from differentiation of totipotent germ cells (3).

The largest series of non germ cell malignancy within germ cell tumours is that of Ulbright *et al* (3) who found 11 cases either in the primary tumour or recurrence on examination of 269 cases of teratoma. The testis was the site of the primary germ cell tumour in 7 of the cases, the mediastinum in 3 and the retroperitoneum in 1. The sarcomatous element of 5 of these patients was rhabdomyosarcoma and 4 of these died of their disease despite Cisplatin based combination chemotherapy. The final patient survived after complete surgical excision. In this group, there was no response to Cisplatin based chemotherapy seen in patients with combined teratoma and rhabdomyosarcoma and where total surgical excision was feasible it was considered the treatment of choice.

Terrier-Lacombe *et al* (4) described a case of rhabdomyosarcoma present within a mature teratoma which developed early metastasis of the purely rhabdomyosarcomatous component despite radical orchidectomy and right lumboaortic lymph node dissection. The patient later fared poorly despite Cisplatin based chemotherapy.

Present management plans for testicular cancer are both standardised and successful and expectations of a favourable outcome are high with over 90% of cases achieving cure (5). Although small in number, men with testicular teratoma containing containing a rhabdomyosarcomatous element would appear to have a poor prognosis by comparison when placed on standard teratoma regimes.

By convention patients with testicular teratoma undergo radical orchidectomy followed by Cisplatin based chemotherapy and restaging in the presence of metastatic spread. Patients with recurrence generally undergo surgical resection followed, if appropriate, by further Cisplatin based chemotherapy. We would recommend that in men with combined teratoma and rhabdomyosarcoma initial management should be as for the teratoma. Remaining or recurring disease requires full histological assessment and in the presence of a significant rhabdomyosarcomatous element, consideration should be given to alternative chemotherapy regimes.

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