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## Abstracts

# Rare malignancies

### ANGIOSARCOMA OF THE RIGHT ATRIUM: A CASE REPORT

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**Background:** Primary cardiac neoplasm occur rarely and most of them are benign. Malignant tumours including angiosarcoma are extremely rare and have a non specific clinical presentation and a poor prognosis. **Case presentation:** We report a case of angiosarcoma of the right atrium presenting on October 2007 progressive exertional dyspnea and edema of extremities. The patient was a 59-year-old woman. Echocardiography, chest computed tomography (CT) revealed a big mass in the right atrium (7 × 6 cm), partially infiltrating the epicardium fat with concomitant pericardial effusion. Moreover, CT abdomen revealed multiple liver metastases. The pt underwent median sternotomy and at the time of exploration, the mass was found in the right atrium with infiltrating the atrioventricular junction. The pathological diagnosis was angiosarcoma. Immunohistochemistry showed the neoplastic cells to be reactive for the endothelial cell markers CD31 and CD34, and negative for the epithelial cell marker cytokeratin, supporting the diagnosis of angiosarcoma. Her performance status was 2 (ECOG scale). The pt agreed to chemotherapy with ifosfamide (3.000 mg/m<sup>2</sup> on day 1, 2, 3) and epirubicin (90 mg/m<sup>2</sup> on day 1), every 3 weeks. After 3 courses of therapy, CT showed a remarkable liver metastases reduction (PR). Further, 3 cycles of chemotherapy was been administered and restaging evaluation imaging revealed only one liver metastase. Treatment is still ongoing and the pt is asymptomatic and alive with disease to seven months since diagnosis. **Conclusion:** The optimum treatment plan is poorly defined in patients with this presentation of angiosarcoma. However, in our experience a good response to chemotherapy treatment was observed.

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### IMATINIB IN THE TREATMENT OF DERMATOFIBROSARCOMA PROTUBERANS

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Dermatofibrosarcoma protuberans (DFSP) is a cutaneous low-grade malignancy with a high recurrence rate and that rarely generates distant metastases. In most cases this tumour is associated with a chromosomal translocation involving the COL1A1 gene on chromosome 17 and the platelet-derived-growth-factor B gene on chromosome 22, generating a fusion gene that constitutively activates the PDGF receptor (PDGFR). In the early stages of disease traditional surgery (wide excision) or Mohs micrographic surgery represents the standard of care. When surgical margins are positive, postoperative radiotherapy is a valuable option. For several decades, treatment of advanced disease has been largely unsuccessful because of the relatively resistance to conventional cytotoxic drugs. Recently it has been shown that inhibiting PDGFR with imatinib can induce a high response rate in case of unresectable or metastatic disease. We saw a young man with a DFSP of the back that underwent tumour resection in 1987. In 1997 a local recurrence was excised without postoperative radiotherapy. The patient came to our attention on January 2004 when the tumour presented secondary lung lesions. On February 2004 the patient underwent a double atypical left lung resection while other smaller lesions were still present. Molecular analysis showed a mutation of the PDGFRb gene. Therefore on July 2004 he started treatment with imatinib 400 mg/day p.o. and a significant PR was observed after just 2 months of treatment. On June 2007 residual lung lesions were resected and the histological examination did not show any malignant cell. The patient is now on treatment and free of disease. Although the treatment of DFSP has significantly improved in the last years, it is likely that a better knowledge of molecular biology will improve response rates and make it possible to discover new effective targeted agents.

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