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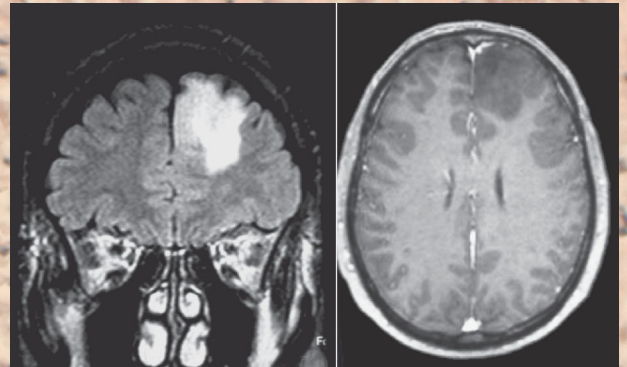
Case Report: Atypical Aggregation of Cancer

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European Association of

NeuroOncology Magazine 2013; 3 (2)

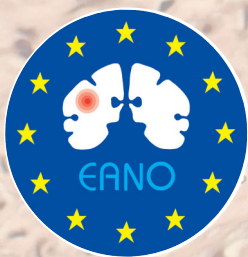
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Atypical Aggregation of Cancer

Emeline Tabouret

From the Department of Neuro-Oncology, AP-HM, Timone Hospital, Marseille, France

■ Case Report

A previously well 36-year-old man presented with moderately severe headache and a progressive, painful, subcutaneous right pectoral swelling. Clinical examination confirmed a pectoral mass, 6 cm of diameter. There were no other general or neurological symptoms or signs. An MRI scan was performed to evaluate the headache (Figure 1) and a CT scan of chest, abdomen, and pelvis to evaluate the chest mass. Brain MRI scan revealed an intrinsic, left frontal neoplasm. Surgical resection of the pectoral mass was performed and histological analysis concluded that the mass was a liposarcoma. A gross total resection of brain tumour demonstrated a WHO grade-II oligo-astrocytoma without 1p19q co-deletion. Methylguanine methyl-transferase (MGMT) promoter methylation status and IDH1 mutation analysis were both negative. The patient received no other treatment for the brain tumour. Two months after brain surgery, the patient was hospitalized unwell and with a fever. Blood count, electrolytes, renal and hepatic functions were all normal. C-reactive protein was elevated at 102 mg/l. Blood cultures were sterile. Viral serology was negative.

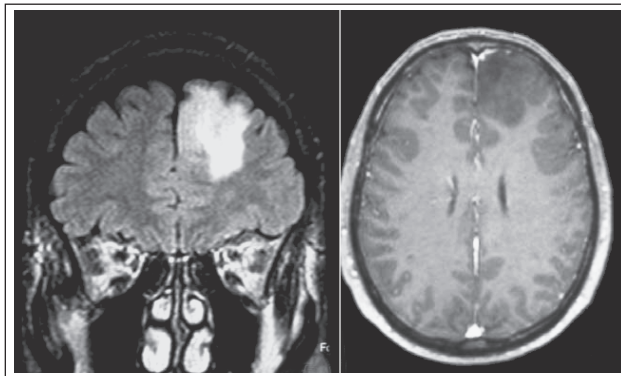


Figure 1. Brain MRI of patient: Flair, T1 with gadolinium and spectrometry analysis.

Analyses of cerebrospinal fluid (CSF) showed an elevated protein at 0.98 g/l. CSF white-cell count was normal and there was no growth of organisms. The CT scan of chest, abdomen, and pelvis also demonstrated a heterogeneous right lung mass and a mass lesion in both adrenal glands (Figure 2).

■ Complete Diagnosis

Pectoral liposarcoma, left frontal low-grade glioma, lung carcinoma with bilateral adrenal metastases in the context of a Li-Fraumeni syndrome.

■ Discussion

Lung biopsy showed a poorly differentiated lung adenocarcinoma. First-line chemotherapy was initiated with cisplatin and vinorelbine. After 3 cycles, the CT-scan showed a progressive disease. Palliative care was initiated. The patient died 2 months later.

The Li-Fraumeni syndrome (LFS) is an autosomal dominant cancer predisposition syndrome associated with soft tissue sarcoma, osteosarcoma, pre-menopausal breast cancer, brain tumours, adrenocortical carcinoma (ACC), and a variety of other neoplasms. More than 70 % of individuals diagnosed clinically have an identifiable disease-causing mutation in tumour suppressor gene p53 (TP53), the only gene known to be associated with LFS. Treatment of clinical manifestations involves routine management of cancers, except for those with breast cancer, where mastectomy is recommended rather than lumpectomy, in order to reduce the risks of a second primary tumour and to avoid radiation therapy. Prevention may include prophylactic mastectomy to reduce the risk of breast cancer in women with a germline TP53 mutation. Prevention of secondary complications includes the avoidance of radiation therapy in order to reduce the risk of radiation-induced

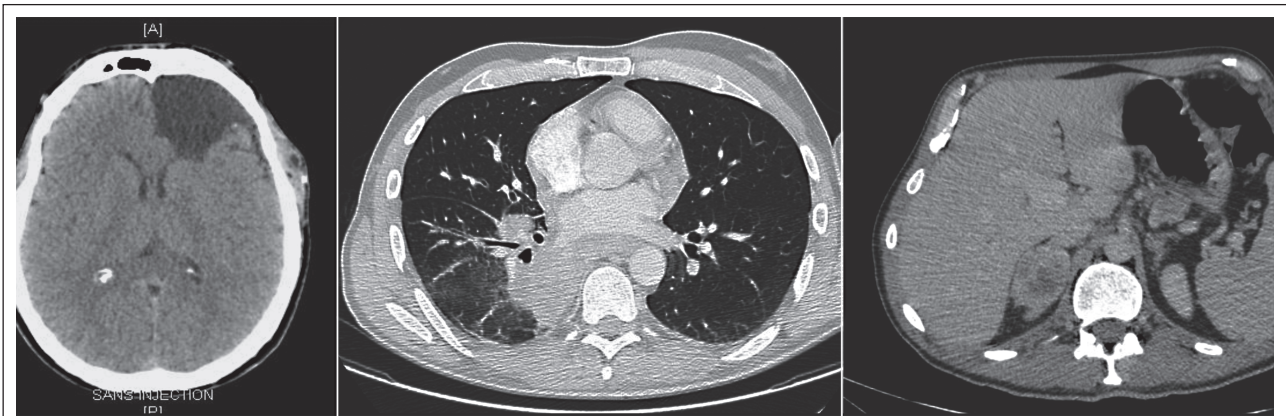


Figure 2. Brain, thoracic, and abdominal CT of the patient.

malignancies. Genetic counselling of relatives who are at risk is appropriate as well as to offer screening to all relatives who are at risk of having a familial TP53 mutation.

Further Reading:

GeneReviews: Li-Fraumeni Syndrome. <http://www.ncbi.nlm.nih.gov/books/NBK1311/> [accessed April 29, 2013].

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Olivier M, Goldgar DE, Sodha N, et al. Li-Fraumeni and related syndromes: correlation

between tumor type, family structure, and TP53 genotype. *Cancer Res* 2003; 63: 6643–50.

Reuss D, von Deimling A. Hereditary tumor syndromes and gliomas. *Recent Results Cancer Res* 2009; 171: 83–102.

Shen L, Sun X, Fu Z, et al. The fundamental role of the p53 pathway in tumor metabolism and its implication in tumor therapy. *Clin Cancer Res* 2012; 18: 1561–7.

Correspondence to:

Emeline Tabouret, MD

Department of Neuro-Oncology, Hôpital de la Timone

264 Rue Saint-Pierre

13385 Marseille Cedex 5

France

e-mail: emeline.tabouret@ gmail.com