Cutaneous angiosarcoma is a rare and aggressive malignant tumor of vascular origin. Multimodality treatment including surgery, radiotherapy and chemotherapy should be used according to age and local spread. Prognosis is poor with a 5-year survival of 10-15%. We report the case of an angiosarcoma of the scalp and face treated with sequential contact radiotherapy and chemotherapy. After local response, the patient progressed in non-irradiated zone then had liver metastasis.

Observation

A 64-year-old man consulted before the rapid increase in the size of a centimeter plate of the scalp in an erythematous and purplish closet, descending on the front, nose and eyelids Figure 1. Histological examination was in favor of an angiosarcoma of high-grade expressing the CD31. The extension work-up completed by a brain MRI and a thoraco-abdomino-pelvic scanner showed the absence of distant metastasis. After a multidisciplinary team meeting, a weekly paclitaxel-based chemotherapy was initiated, associated with palliative radiotherapy by contact (150Kv) issued at a dose of 40 Gy on the frontal lesion, which was the most extensive and the thickest. The evolution was marked by a net improvement of the irradiated lesion that has become depigmented and flat Figure 2. A local progression was observed for the other lesions that have afterwards been irradiated Figure 3. The patient progressed locally outside the treated fields Figure 4. Three months later, he consulted with dyspnea and a deteriorated general condition. The updating of the extension examination was in favor of multiple liver metastases and the presence of a pneumothorax that was drained in emergency. Before the metastatic dissemination and the bad general state, therapeutic abstention was retained. The patient died 15 days after leaving the hospital.
Heterogeneous symptoms, sometimes simulating benign lesions in type of bruising or hemangiomas can drag out the diagnosis and make any resection difficult, extensive and mutilating. Most angiosarcomas occur spontaneously but malignant transformations of benign vascular lesions have been described in the literature [7]. Several risk factors are associated with angiosarcomas. Chronic lymphoedema known as Stewart–Treves syndrome, arising after surgery, radiotherapy or as a part of Milroy’s disease is the most common one. The radiotherapy also constitutes an independent risk factor. Moreover, exposure to chemicals such as arsenic, vinyl chloride and thorium dioxide increases the risk of developing an angiosarcoma [3].

The role of immunosuppression in the pathogenesis of angiosarcomas is undetermined. Several cases following a renal transplant have been reported [8]. Goedert et al. suggested that similarly to Kaposi sarcoma, angiosarcoma and AIDS could be associated [9]. Although the role of Human Herpes Virus 8 (HHV8) in Kaposi sarcoma is proven, its involvement in the pathogenesis of angiosarcomas was not found [10].

The immunohistochemistry plays an important role in the diagnosis of the angiosarcoma and especially for those that are undifferentiated. Ulex lectin and CD31 are more sensitive and specific endothelial cell markers of skin angiosarcoma [11].

Angiosarcoma is an aggressive tumor, with extended skin dissemination, relapsing locally and giving early metastases through blood and/or lymphatic. Metastases are most often of lung, bone, lymph node and liver. The lung is the most common site and the leading cause of death. In a Japanese autopsy series of 95 patients [12], pulmonary metastases were found in nearly 70% of cases.

Besides, primary tumors located at the scalp level had a more frequent rate of pulmonary complications, type of pneumonia, pneumothorax and hemothorax, than the other locations [12]. In a more recent study of 37 cases, which focused only on angiosarcoma of the scalp and face, lung metastases occurred in nearly 77% of patients [13].

The prognosis for cutaneous angiosarcoma of the scalp and face remains dark with 5-year survival rates in older series of 10–15% [1,2,14]. More recent studies, although retrospective, reported survival rates at 5 years from 38 to 54% thanks to improved techniques of radiotherapy and chemotherapy [15–18] Table 1. In addition, patients of less than 70 years, more inclined to tolerate a multimodal approach to their treatment (surgery, radiotherapy, chemotherapy), seem to have less local recurrence and better survival [6,18,19].

Surgical treatment remains the cornerstone of treatment even though obtaining a complete surgical excision is difficult in front of the local extent at the scalp and face area [20].

The use of radiation therapy associated to surgery allows a better local control of the disease [9,15,20,21]. Recommended doses appear to be 70 Gy or more in case of non-operated tumor
Table 1: Survival rates of angiosarcoma of scalp and face in literature.

| Author (year)                      | Number of patients | Therapeutical modalities | OS (%) | MST (months) |
|-----------------------------------|--------------------|--------------------------|--------|--------------|
| Hodgkinson et al. (1979) [1]      | 13                 | RT / S                   | 15 NA  | NA           |
| Holden et al. (1987) [2]          | 72                 | RT / S                   | 12 NA  | NA           |
| Ohguri et al. (2005) [18]         | 20                 | RT / S / CT / rIL-2      | NA     | 36.2         |
| Guadagnolo et al. (2011) [19]     | 70                 | RT / S / CT              | 43     | 41           |
| Ogawa et al. (2012) [13]          | 48                 | RT / S / CT / rIL-2      | NA     | 13.4         |
| Miki et al. (2013) [19]           | 17                 | RT / S / CT / rIL-2      | NA     | 26           |
| Dettenborn et al. (2014) [16]     | 80                 | RT / S / CT              | 54     | 64           |
| Patel et al. (2015) [17]          | 55                 | RT / S / CT              | 38     | 25.2         |

Abbreviations: RT = Radiotherapy; rIL-2 = recombinant Interleukin 2; S = Surgery; CT = Chemotherapy; OS = Overall Survival at 5 years; MST = Median Survival Time; NA= Not Available.

1 In this study, only patients with angiosarcoma of the scalp were included.

The rarity of this pathology makes phase II/III studies difficult to undertake and the best therapeutic sequence between the different available methods involving surgery, radiotherapy, immunotherapy and chemotherapy, still need to be defined.

Conclusion

The angiosarcoma of the scalp and face is a rare pathology. Its local aggressiveness and its metastatic dissemination (notably at the pulmonary level) make a very bad prognosis tumor. The young age, the extent of the lesions and the quality of surgical excision are the main prognostic factors. A multimodal therapeutic approach seems to offer the best chances of patients’ survival.

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