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ORIGINAL ARTICLE

Cutaneous Angiosarcoma: Clinical and Pathology Study of 16 Cases☆

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Abstract

Background and objectives: Primary cutaneous angiosarcoma is one of the most aggressive skin tumors and carries a very poor prognosis. Its initially indolent clinical presentation explains the frequently late diagnosis that, together with its typically multifocal pattern and poor delimitation, often makes surgery difficult. The low incidence of primary cutaneous angiosarcoma means that few large single-center series have been published. We review the clinical and pathologic characteristics of cutaneous angiosarcomas treated in our hospital, looking for prognostic factors and for possible diagnostic traits that could facilitate early diagnosis.

Material and methods: This was a retrospective observational study including all patients diagnosed with cutaneous angiosarcoma in Instituto Valenciano de Oncología in Valencia, Spain between January 2000 and December 2015. We recorded 16 clinical parameters, including age, sex, type of angiosarcoma, site, size, and time since diagnosis, and 8 histopathologic parameters.

Results: We identified 16 patients (11 women and 5 men) with cutaneous angiosarcoma. Their mean age was 67 years (median, 71 years). The most common site was the trunk (10 cases), followed by the head and neck (5 cases). The mean size of the tumor was 10 cm (median, 6.5 cm). Fourteen patients underwent surgical excision. Six of the 16 patients were alive at the end of the study, after a mean follow-up period of 42.5 months.

Conclusions: The major determinants of survival among patients with cutaneous angiosarcoma are tumor size and patient age. Other characteristics associated with a poor prognosis were infiltration of deep planes (muscle), a predominantly solid histologic pattern, and a larger number of mitoses.

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PALABRAS CLAVE
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Factores pronósticos

Angiosarcoma cutáneo: estudio clínico-patológico de 16 casos

Resumen

Introducción y objetivos: El angiosarcoma primario cutáneo es uno de los tumores más agresivos y de peor pronóstico de la piel. Su clínica inicialmente indolente justifica frecuentes diagnósticos tardíos, lo que sumado a su carácter muchas veces multifocal y a su mala delimitación suele dificultar la cirugía. Debido a su baja frecuencia existen pocas series largas de casos tratados en un mismo centro. Revisamos las características clínico-patológicas de los angiosarcomas cutáneos tratados en nuestro centro en búsqueda de factores pronósticos, así como de posibles rasgos que faciliten un diagnóstico precoz.

Material y métodos: Se realizó un estudio observacional retrospectivo de todos los pacientes diagnosticados de angiosarcoma cutáneo atendidos en nuestro hospital entre enero de 2000 y diciembre de 2015. Se recogieron 16 parámetros clínicos incluidos —entre otros—edad, sexo, tipo de angiosarcoma, localización, tamaño, tiempo de evolución y además 8 parámetros histopatológicos.

Resultados: Se recogieron 16 pacientes con angiosarcoma cutáneo —11 mujeres y 5 varones—, la media de edad fue de 67 años y la mediana de 71 años. La localización más frecuente fue el tronco con 10 casos, seguida de la cabeza y el cuello con 5 casos. La media del tamaño tumoral fue de 10 cm y la mediana de 6,5 cm. Se realizó escisión quirúrgica del tumor a 14 pacientes. Tras una media de seguimiento de 42,5 meses, 6 de los 16 pacientes seguían vivos al finalizar el estudio.

Conclusiones: La supervivencia de los pacientes con angiosarcoma cutáneo viene determinada principalmente por el tamaño tumoral y la edad. Otros rasgos asociados a peor pronóstico en nuestros pacientes fueron la infiltración a planos más profundos (músculo), un patrón histológico predominantemente sólido y un mayor número de mitosis.

Cutaneous angiosarcoma has one of the worst prognoses among all cutaneous tumors. It is very aggressive and has high local recurrence rates. The 5-year survival rate is between 12% and 34% according to most studies,[1,2] but it can be as high as 62%.[3] Unlike other sarcomas, degree of differentiation has not been associated with prognosis in cutaneous angiosarcoma.[4]

The classic form of cutaneous angiosarcoma is an ill-defined bruise-like edematous lesion with a largely indolent clinical presentation in its early phases. It occurs on the face or scalp of elderly patients (Wilson-Jones angiosarcoma) and accounts for approximately 50% of all primary cutaneous angiosarcomas.[5–8] Two other typical forms of angiosarcoma are Stewart-Treves syndrome, which develops in areas of long-standing lymphedema and is particularly common in women who have undergone radical mastectomy,[9–11] and postradiation angiosarcoma, which develops in areas of irradiated skin, particularly in the pectoral region of women with a history of breast cancer treated with radiation therapy.[12–15]

The histopathologic appearance of cutaneous angiosarcoma varies from relatively differentiated forms with recognizable vascular spaces covered by prominent endothelial cells with some atypia and an infiltrative pattern dissecting the collagen bundles to more solid highly undifferentiated forms composed of spindle or epithelioid cells with considerably greater atypia and pleomorphism and a higher number of mitoses. Vascular spaces are rare and the tumors can sometimes mimic carcinoma.

The mainstay treatment for cutaneous angiosarcoma—and the only one that is potentially curative if disease-free margins are achieved—is surgical excision with wide margins followed by local radiation therapy and even, in the opinion of some authors, radiation of regional lymph nodes.[6] In most cases, however, it is not easy to achieve disease-free margins due to extensive subclinical spread. In addition, these tumors are frequently multifocal. Chemotherapy has a purely palliative role in the management of cutaneous angiosarcoma.

Although cutaneous angiosarcoma is rare (accounting for less than 1% of all sarcomas), most cases of angiosarcoma originate in the skin. Due to their low prevalence, cutaneous angiosarcomas are included in series of visceral or bone angiosarcoma, which have an even more dismal prognosis.[1] There are thus few large series of cutaneous angiosarcoma in the literature due to the shortage of long-term, uniform cases.[2,4,5,7,16] Management of angiosarcoma is additionally often disheartening, above all in cases of advanced disease, which have a very poor prognosis despite the use of aggressive treatment from the outset. Motivated thus by the difficulties associated with the management of cutaneous angiosarcoma and the little literature available, we investigated all cases of cutaneous angiosarcoma treated at Instituto Valenciano de Oncología (IVO), in Valencia, Spain, with the aim of identifying clinical, histologic, and treatment-related factors possibly associated with prognosis. To do this, we reviewed medical records and clinical findings in search of exploratory data that could serve as