Adult Rhabdomyoma

12

Definition

Adult rhabdomyoma is a benign tumor showing mature skeletal (striated) muscle differentiation. It is also known as adult extra-cardiac rhabdomyoma, rhabdomyomatous hamartoma, and rhabdomyoma purum.

Along with fetal rhabdomyoma and genital rhabdomyoma (→ see dedicated sections), it belongs to the extra-cardiac subgroup of rhabdomyomas (→ see also the section entitled "Rhabdomyoma").

Epidemiology and Presentation

It is a very rare neoplasm representing <2% of all muscular tumors. Most cases develop in the head and neck (parapharyngeal space, salivary glands, larynx, mouth, and soft tissue of the neck). Very rarely it has been described in other sites such as the mediastinum. The lesion occurs mainly in men (M:F = 3:1), the median age being 60 years (range: 30–80 years). The tumor commonly presents as a slow-growing, painless mass (median diameter: 3 cm) which can cause compression of adjacent structures. The tumor is multifocal in 25% of cases.

Unlike cardiac rhabdomyoma and fetal rhabdomyoma (→ see dedicated sections), adult rhabdomyoma is not associated with either basal cell nevus syndrome (Gorlin syndrome) or with tuberous sclerosis complex (TSC).

Pathology

Tumor cells are large and polygonal with abundant granular eosinophilic cytoplasm and small round vesicular nuclei; the cytoplasm may be vacuolated ("spider cells") or contain rod-like inclusions or cross striations (well-differentiated skeletal muscle cells). Neither mitotic activity nor atypia are encountered.

S. Mocellin, *Soft Tissue Tumors*, https://doi.org/10.1007/978-3-030-58710-9_12

Differential diagnosis may be needed with the following: alveolar soft part sarcoma, granular cell tumor (no skeletal muscle differentiation, smaller cells with poorly defined cell borders, often overlying pseudoepitheliomatous hyperplasia, S100 positive); hibernoma (no skeletal muscle differentiation); paraganglioma (positive for NSE,[1] synaptophysin, and chromogranin); and well-differentiated rhabdomyosarcoma.

Biomarkers

Adult rhabdomyoma stains positive for MSA,[2] desmin, and myogenin, whereas it stains negative for EMA[3] and CD68.

Prognosis

Adult rhabdomyoma is a benign tumor, but it often recurs. However, it does not infiltrate contiguous structures or metastasize.

Therapy

Surgical excision is the treatment of choice.

[1] NSE: neuron-specific enolase.

[2] MSA: muscle-specific actin.

[3] EMA: epithelial membrane antigen.