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Rhabdomyomatous Mesenchymal Hamartoma – Case Report

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Abstract

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Introduction

Rhabdomyomatous mesenchymal hamartoma of skin is an extremely rare lesion of the dermis and soft tissue. Grossly, most lesions are polypoid and attached to the skin by a long stalk, with circumferential constriction of the distal attachment site. Histologically, single or small groups of mature-appearing skeletal muscle fibers are found within the subcutaneous tissue and dermis. The fibers frequently are deposited in a collagenous stroma admixed with mature adipose tissue and adnexal structures. Blood vessels and nerves may also be found admixed among the mature skeletal muscle fibers.

Case Report

A 40 year old paraplegic patient presented with ulcer and exophytic lesion in the gluteal region of 1 month duration, with a clinical suspicion of squamous cell carcinoma. MRI of the patient revealed mesenchymal tumor with likely malignant transformation.

Gross

Received a globular tissue mass with elliptical piece of skin, whole soft tissue mass measuring 12 x 9 x 5, surface of skin showed pedunculated mass with pedicle measuring 4x 3x2.5cm. c/s grey white (Fig 1 & 2).

Figure 1

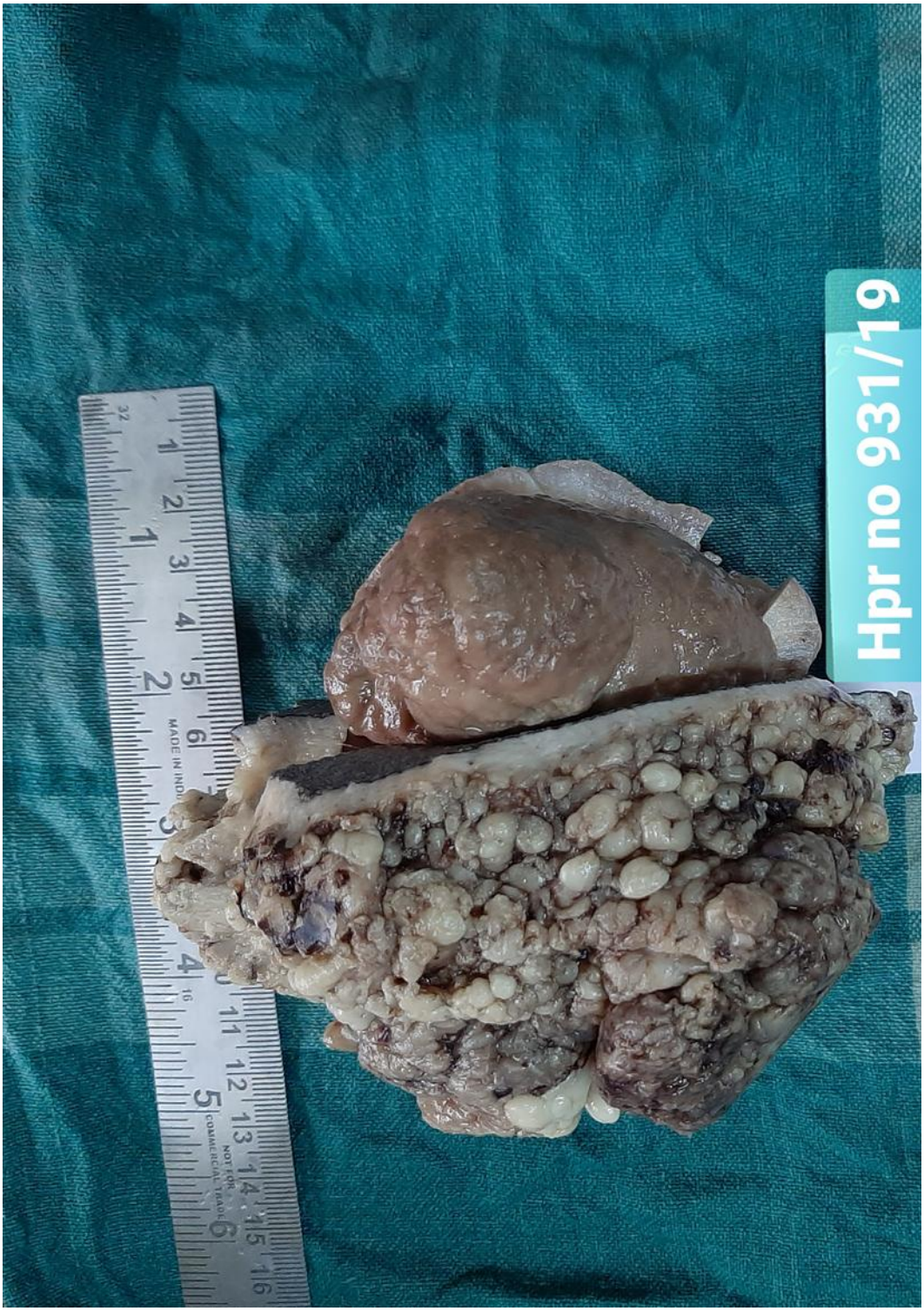

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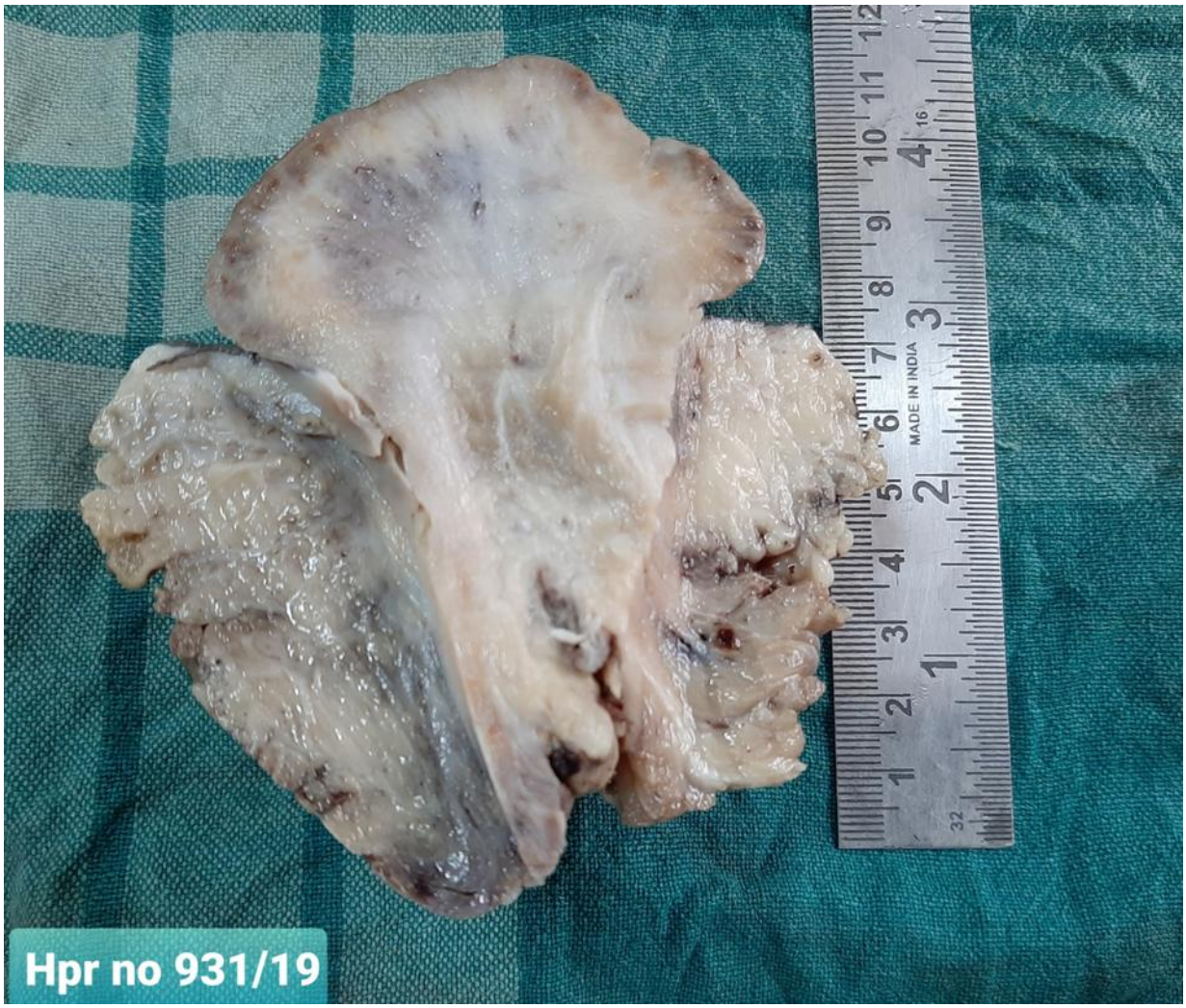
Figure 2



Microscopy

Section shows bundles of bland spindle shape cells intervened by collagen bundles, extensive myxoid areas and numerous dilated vascular channels. Deeper areas of lesion showed skeletal muscle differentiation. Surrounding fat and muscle tissue shows extensive fibrosis (Fig 3).

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Figure 3

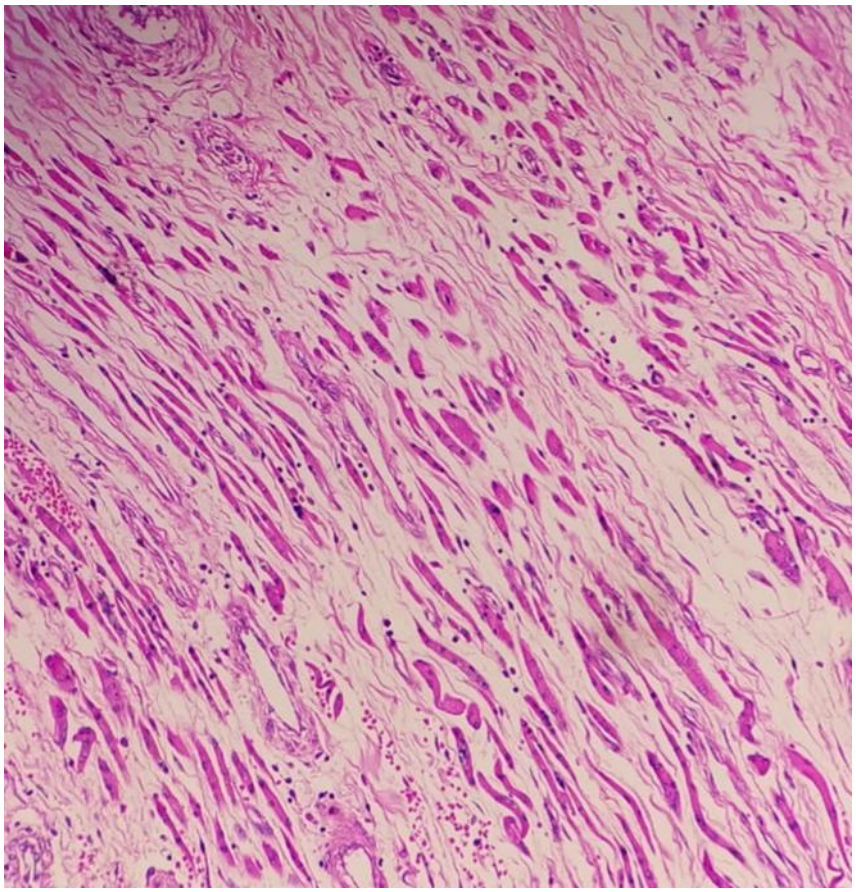

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Figure 4

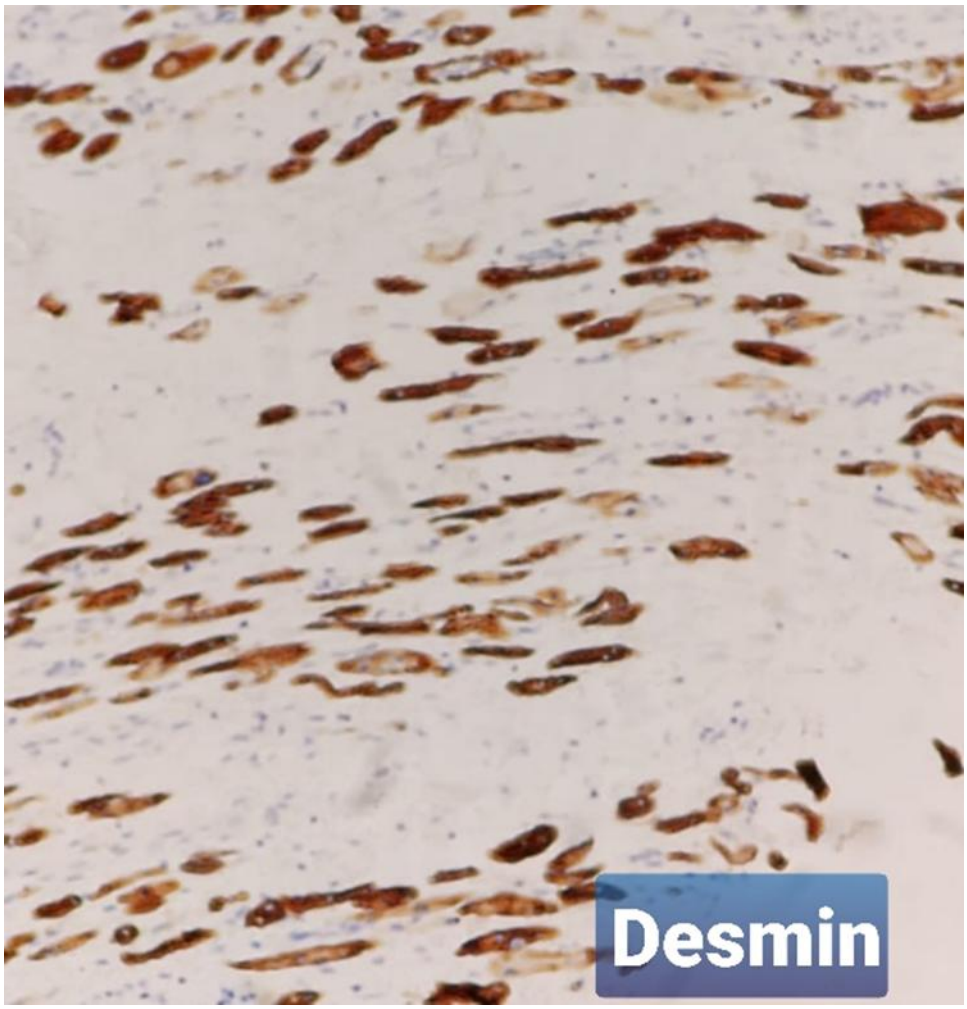

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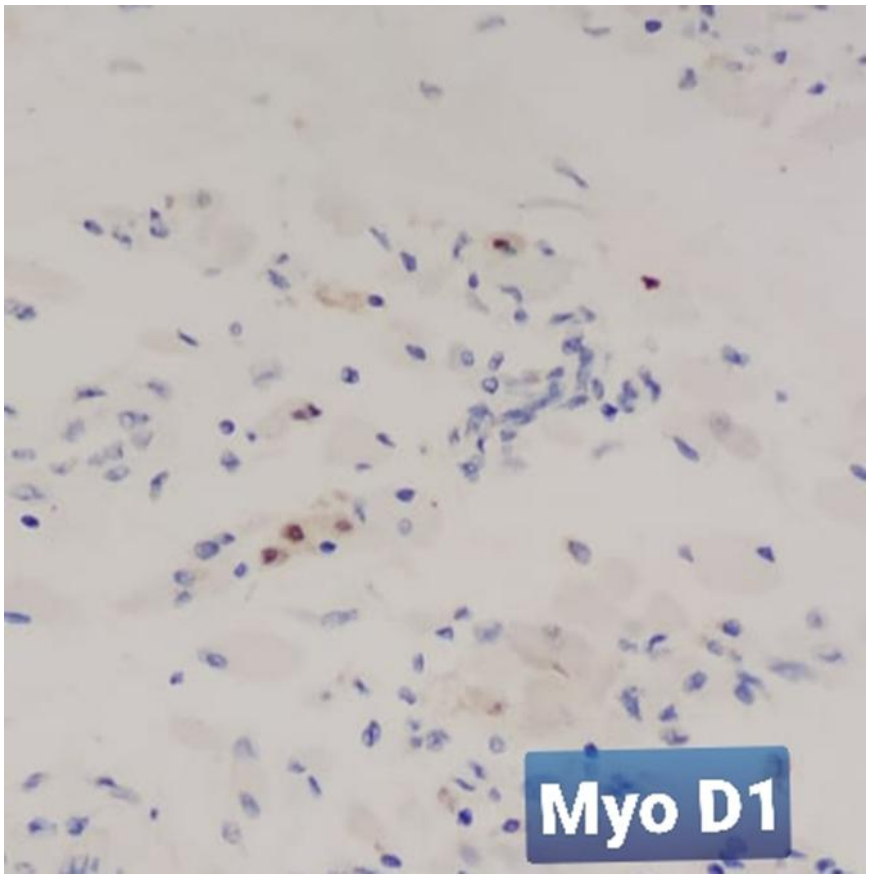
Fig 4 shows Desmin positivity and Fig 5 shows scattered Myo-D 1 positivity.



Discussion

Rhabdomyomatous mesenchymal hamartoma typically presents as a small dome-shaped papule or a polypoid pedunculated lesion.Most common location appears to be the chin, followed by the periorbital, periauricular and anterior mid-neck region.Incidence of this lesion is more in newborns but cases has been reported in adults also.This lesion is adequately treated by local excision, and recurrences have not been described.

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