A 40-year-old male patient referred with a chief complaint of swelling in the right cheek for 5 years. The swelling showed a gradual increase in size for over 6 months. No other relevant history in terms of habit, medication or systemic disease was given by the patient. On extra-oral examination (Figure 1a), mild swelling was observed in the cheek region that extended posteriorly towards the angle of the mandible. The skin over the swelling was normal and no other abnormalities were noted. Intra-oral examination showed no visible swelling or abnormality. On palpation, the swelling was firm and mobile with no distinct edges. Swelling was noted in the masseter muscle. For the same complaint, the patient had visited a regional diagnostic center about 5 months ago, where fine needle aspiration cytology (FNAC) had been performed. The findings of aspiration cytology showed hemorrhage and gave the impression of a vascular lesion. Further, ultrasonography (USG) and magnetic resonance imaging (MRI) were done to confirm the findings of FNAC. USG and MRI showed a lesion in the right masseter muscle measuring approximately 2 × 1.7 × 2.3 cm with a few specks of calcification. The lesion was in contact with, but not extending into, the superficial part of the right parotid gland. The parotid and submandibular salivary glands appeared normal. The MRI showed iso/hyper-intense signals on T1-wt images and hyper-intense signals on T2-wt/Stir images (Figure 1b). The overall impression was of a mixed-signal intensity vascular lesion in the right masseter muscle with possible calcification. Further, an excisional biopsy was performed and a single mass of tissue was obtained which was soft to firm in consistency, grayish-brown in color. It was well-circumscribed with a pebbly surface measuring about 4 × 3.3 cm. Histopathological evaluation of the lesion showed the longitudinal and transverse section of muscle bundles with interspersed endothelial-lined, dilated large and medium-sized blood vessels (Figures 2a and 2b). A few lymphatic channels and adipocytes were also noted alongside the fibrin filled blood vessels & muscle bundles (Figure 3a). Areas of thick and thin-walled arteries engorged with red blood cells were noted (Figure 3b). Deeper section showed focal calcification.

What is your diagnosis?
See the next page for your diagnosis.
Intramuscular hemangioma (IMH) accounts for less than 1% of all soft tissue hemangiomata. However, in the head and neck, massteric IMH is the most common and usually presents as a slow-growing swelling. IMH is known to occur in the first three decades of life while our case presented in the 4th decade. Two key theories have been put forth concerning the etiogenesis of IMH, the first being repetitive trauma and the second, due to hormonal influence. However, IMH is considered a congenital lesion made of embryonic sequestration similar to arteriovenous malformation.

Histologically, IMH presents with capillary and/or cavernous spaces that infiltrate the skeletal muscle and can be accompanied by adipose tissue. It has been classified into cavernous (large vessels >140 mm diameter), capillary (small vessels <140 mm diameter) and mixed type IMH. The mixed subtype has a documented local recurrence of 28% while the cavernous IMH shows recurrence of only about 9%. Additionally, phleboliths or calcifications are noted in approximately 15%-25% of IMH. Clinically, the presence of calcification or IMH itself presents with negligible to minimal signs and symptoms. The turkey wattle sign is one of the few signs observed in IMH, wherein the lesion enlarges with clenching of the teeth or in a specific head position. A change in the venous pressure is implicated as the reason for this transient size change.

Overall, the location, rarity of occurrence and lack of apparent symptoms cause difficulty in diagnosis of IMH. Various treatment modalities have been suggested for IMH based on the size and location of the lesion. Treatment modalities commonly employed include cryotherapy, lasers, use of sclerosis agents, steroid treatment, radiation therapy and complete surgical excision. As the extent of the lesion cannot be traced in most cases, complete surgical removal of the muscle is advocated. These further aids in minimizing the recurrence that could occur with incomplete removal of the lesions tissue. In the present scenario, the patient was provided with information regarding the condition, possible treatment options and associated outcomes. On consenting for surgical excision, removal of the lesion was accomplished and the patient was recurrence-free after one year.

Authors’ Contribution
Images and data: AR, KMD and PVA; Manuscript draft: KMD and ADK; Revision and final manuscript preparation: KMD and PVA.

Conflict of Interest Disclosures
None.

Ethical Statement
Informed consent was obtained from the patient for use of photographs and for publication of this case.

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References