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EP01

UNVEILING A RARITY: A CASE STUDY OF DERMATOFIBROSARCOMA PROTUBERANCE MANIFESTATION, DIAGNOSIS, AND THERAPEUTIC MANAGEMENT

M. A. Ilani Ismail[1], M. A. Kamarudin[1], H. K. Gnanasegaram[2]
[1]Department of Radiology, Hospital Kuala Lumpur, Jalan Pahang, 50586 Kuala Lumpur, Malaysia
[2]Department of Pathology, Hospital Kuala Lumpur, Jalan Pahang, 50586 Kuala Lumpur, Malaysia

Abstract: Dermatofibrosarcoma protuberance (DFSP) is a rare soft tissue tumour typically presented in young adult as a slow growing and a firm plaque. However, few patients may have accelerated growth, painful swelling or even in rare cases ulceration, and bleeding in case of large tumour. We report a case of bleeding DFSP in a 27-year-old gentleman with a rapid growing left clavicular lump for 9 months, complicated with bleeding from left chest wall and associated stabbing pain. His MRI reveal a superficial subcutaneous avidly enhancing soft tissue tumour multiple flow void within. Patient underwent pre-operative angioembolisation showing tumour blush from clavicular and pectoral branch of left thoracoacromial artery which was later embolized. Subsequent surgical resection was done with no complication perioperatively. HPE later confirm the diagnosis of Dermatofibrosarcoma protuberance with immunohistochemistry shows spindle cell was positive for CD34. We also discuss the clinical and imaging features, as well as the treatment modalities for DFSP in particular preoperative tumour embolization.

Introduction: Dermatofibrosarcoma protuberance (DFSP) is a low-grade, relatively uncommon, soft-tissue sarcoma that develops in the skin's connective tissue of young adult [1] It was originally described in 1924 by Darier and Ferrand. The term dermatofibrosarcoma protuberans was coined by Hoffman in 1925 to reflect its origin in the skin's connective tissue and its tendency to protrude from the skin [2]. It typically appears as a firm, raised nodule or plaque on the skin that is painless and slow growing. DFSP is mostly found on the trunk, limbs, and head and neck regions of the body. Diagnosis of DFSP typically involves a physical examination of the skin lesion, followed by a biopsy to confirm the diagnosis [3]. Radiological appearance of this tumor has rarely been studied and findings infrequently discussed in the literature as many lesions underwent resection before imaging. However, imaging does play a vital role in the differentiation of this tumor in order to differentiate this from other aggressive soft tissue tumor such as more aggressive sarcomas and hemangioma [1]. While most of DFSP present as plaque, few cases present with bleeding. Bleeding DFSP can be particularly challenging to manage, as it can be difficult to achieve haemostasis during surgery or even during biopsy. We report a case of bleeding DFSP of which pre-operative embolization was performed to ensure good surgical and patient outcome.

Material and Method: We report a case of DFSP in a 27-year-old gentleman with no significant previous medical illness. He initially presented with a rapid growing left clavicular lump for 9 months which started as a small lump, then rapidly increasing in size within 1 month prior to presentation. It is associated with bleeding from the left chest wall and stabbing pain which require multiple admissions. Physical examination reveal a mass over left upper anterior chest wall with bleeding abrasion wound on the surface of the lumps. MRI of the tumour reveal a superficial subcutaneous soft tissue tumour which demonstrates low signal intensity on T1WI, intermediate to high signal on T2WI and avid enhancement on post contrast study with multiple flow void within. The mass abuts the left pectoralis muscle without frank invasion with clear plane between the mass and clavicle, left subclavian vein and arteries. Neurovascular bundle is preserved. CT angiogram (CTA) later showed the arterial feeder from to the mass arising from superior thoracic artery and left thoracoacromial branch of the left subclavian artery. Venous phase of the CTA demonstrates multiple subcutaneous venous varix surrounding the inferomedial aspect of the mass. We performed a transfemoral angiogram of the left subclavian artery and selectively cannulate the branches of the left subclavian artery which were the thyrocervical, dorsal scapula, superior thoracic, lateral thoracic and thoracoacromial artery. The main feeder of the tumour arises from the thoracoacromial artery which branches into the clavicular and pectoral branch. These arteries were embolized using 355-500 polyvinyl alcohol (PVA) particle. Another feeder from the acromial branch was not embolize as it is too tortuous to navigate and has a risk of non-targeted embolization towards the axillary artery and the left upper limb. 24 hours post angioembolization, patient proceeded with wide excision of the tumour and closure of the defect with split thickness skin graft (harvested from patient's right thigh).

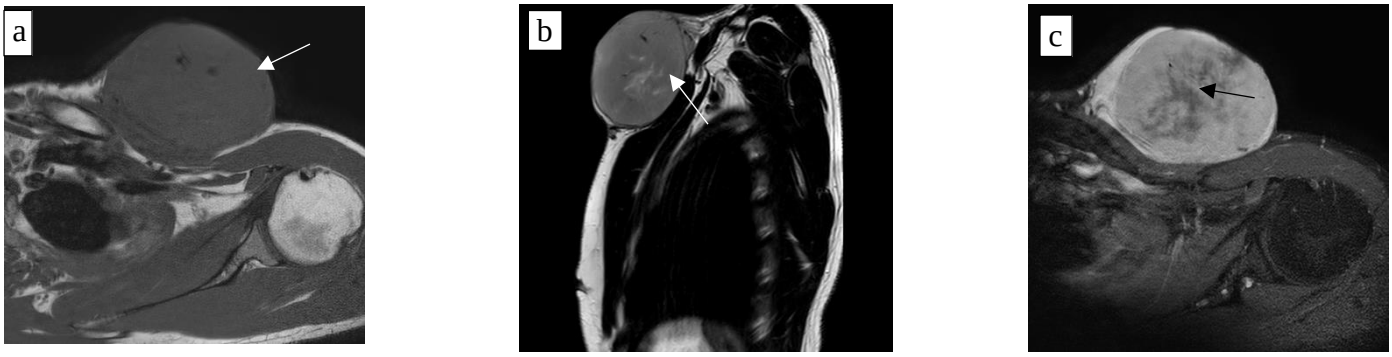

Figure 1

Figure 1: Axial (**a** and **c**) and sagittal (**b**) MR images of anterior chest wall mass which demonstrates hypointense signal on T1WI, intermediate to hyperintense signal on T2WI (*white arrows*) and avid enhancement post contrast with multiple flow void within (*black arrow*).

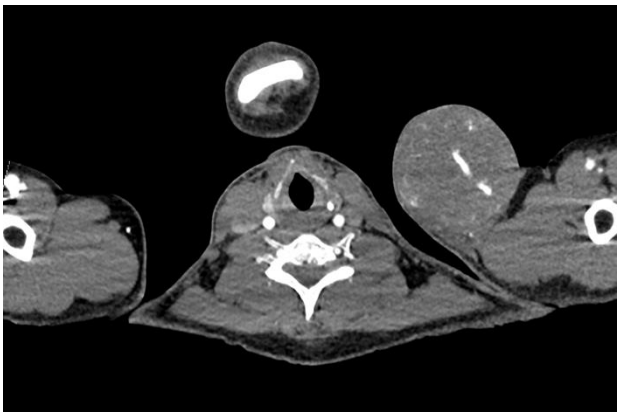

Figure 2: CT angiogram showing arterial feeder suppling the tumour at left upper anterior chest wall (*arrow*).

Results: Prior to angioembolization and wide excision of the tumour, patient's haemoglobin show slight reducing in trend on the day of admission from 14.3g/dl (21.5.2023) to 12.6g/dl (22.5.2023). Post operatively, patient's haemoglobin did not show reducing trend and increased to 13.0g/dl (23.5.2023). Patient's pain was well-controlled, hemodynamically stable and was on antibiotic coverage for the skin graft that is continued until patient discharged to plastic surgery ward on 23.5.2023. Patient was discharged from hospital a week later and recovered well. HPE of the tissue subsequently confirmed the diagnosis of DFSP with immunohistochemistry showing spindle cell was positive for CD34. The resection margin of this tumour shows adequate margin clearance.

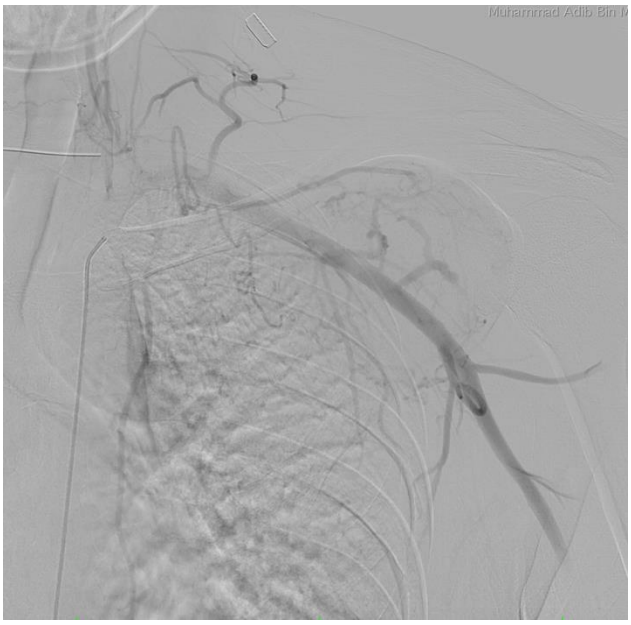

Figure 3: Pre embolization run showing arterial feeder to the tumour.

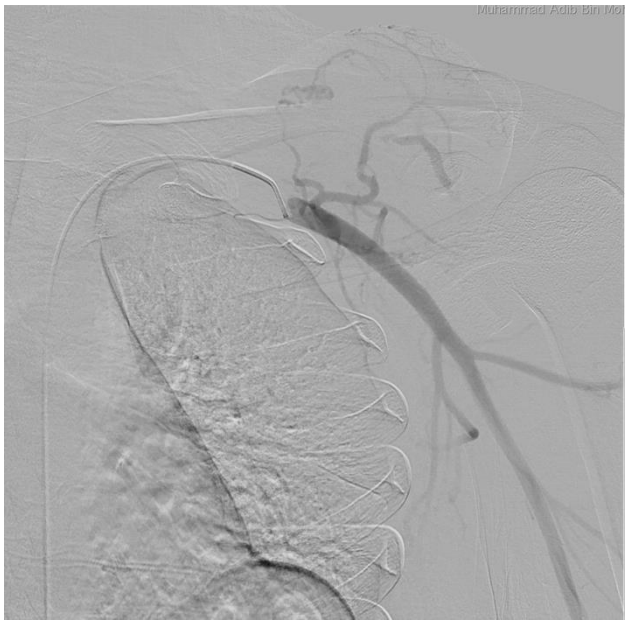

Figure 4: Post embolization run showing reduced arterial feeder supplying the tumour.

Discussion: DFSP is a rare soft tissue tumour and males are slightly more commonly affected with the highest frequency reported between the second and the fifth decades [4,6]. The trunk appears to be the most common site followed by the extremities and the head and neck region [6]. Patients often ignore this tumor due to their slow growth and usually present when tumour has already large enough. Hence, few number of case reports discussed on the early and late imaging appearance of this entity. Local recurrence is a major concern after surgical excision. The frequencies of local recurrences ranged from 20% to 50% [3]. Thus, a proper excision with good safety margin is important in order to reduce the risk of recurrence in this patient. In our case, resection margin of the tumour was adequate for margin clearance. A bleeding DFSP, which one the complication to happen when there are abundant blood supply supplying it, is difficult to manage. Pre-operative embolization of the tumoral mass may help to reduce its size and facilitating the surgical act with minimal bleeding [5]. In our case, patient initially had a reducing haemoglobin trend with anaemic symptoms contributed by multiple bleeding episodes. We applied our expertise in doing embolization of the arteries supplying the tumour which has helped the surgeon to control haemostasis during the operation and easier resection of the tumour.

Conclusion: Bleeding DFSP is challenging to manage and requires a multi-disciplinary team approach. Pre-operative embolization yield a good surgical and patient outcome.

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EP02

MANAGEMENT AND OUTCOMES OF CAROTID CAVERNOUS FISTULAS: A RETROSPECTIVE ANALYSIS OF HOSPITAL KUALA LUMPUR EXPERIENCE

Z. Ismail[1], M. A. Kamarudin[1]
[1]Department of Radiology, Hospital Kuala Lumpur, Jalan Pahang, 50586 Kuala Lumpur, Malaysia

Introduction: Carotid cavernous fistulas (CCF) are abnormal vascular shunts between the carotid arterial circulation with its branches and the cavernous sinus venous system. In the past, conservative management was somewhat an effective treatment alternative for CCF however recently, multiple endovascular therapy (EVT) of CCF with a variety of embolization material using different approaches have enabled higher occlusion rates with good clinical and functional outcomes. The main purpose of this study is to report the treatment outcomes of CCF using EVT with different neuroendovascular techniques at Hospital Kuala Lumpur (HKL), a tertiary government hospital in Malaysia.

Materials and Methods: This is a retrospective study over a period of 36 months (year 2020 to 2023) at HKL, which assessed the treatment outcomes of patient who underwent EVT of CCF. Patient records and all imaging data including angiograms were reviewed for demographic and epidemiological data, symptoms, fistula type, number of EVTs, complication of EVT, type of embolic materials, occlusion rates and recurrences.

Results: A total of 52 patients between 13 years old and 81 years old were treated in our study. Of these, the aetiology of 31 patients (59.6%) were spontaneous, 19 patients were post-traumatic (36.5%) and the remaining 2 patients (3.8%) were due to ruptured cavernous aneurysms. A majority (76.9%, 40/52) of the cases were completed in one session, with 8 patients (15.3%) needing to undergo additional treatment. The most frequently used access was that of combined (51.9%, 27/52), followed by transarterial (32.7%, 17/52) and transvenous catheterization (15.4%, 8/52). Exclusively coils were used in 34.6% (18/52), and a combination of glue and coils in 21.2% (11/52). Complete obliteration was achieved in 86.5% of patients (45/52) with an intraprocedural-related complication rate of 7.7% (4/52) and no mortality.

Conclusion: Elevated curative response with low rates of morbidity in relation to intraprocedural complications using EVT for CCF have shown safe, positive and effective outcomes, even in complex scenarios.

EP03

HYPERVASCULAR MELANOTIC NEUROECTODERMAL TUMOR OF INFANCY IN THE MAXILLARY REGION SUCCESSFULLY TREATED WITH SUPERSELECTIVE EMBOLIZATION AND EXCISION: A CASE REPORT

D. Andriawan[1], Trianingsih[2], A. Rahman[2]

[1]Faculty of Medicine, Public Health and Nursing Universitas Gadjah Mada, Daerah Istimewa Yogyakarta 55281, Indonesia

[2]Department of Radiology, Dr. Sardjito General Hospital, Daerah Istimewa Yogyakarta 55281, Indonesia

Introduction: Melanotic Neuroectodermal Tumor of Infancy (MNTI) is a very rare benign neoplasm of neural crest origin occurring in infants mainly during the first year of life. Hypervascularization is an unusual feature of MNTI. Treatment is primarily done by surgical excision. Angiography and embolization are rarely performed but could be an option if there is a risk of severe bleeding during surgical intervention. To the best of our knowledge, no prior studies have described preoperative angiography and embolization for hypervascular MNTI in the maxillary region.

Case Report: A 6-month-old male infant was referred from previous hospital with a chief complaint of a mass in the upper gum that progressively increased in size. The mass was initially suspected of being a hemangioma and the infant was referred to our hospital for surgical treatment. Head MRI using T1W, T2W, FLAIR, DWI / ADC, SWI, 3D-CE T1W, and 3D-T1-MPRAGE sequences revealed a mass in the left hard palate that was vascularized by branches of the left maxillary artery and shows imaging features similar to hemangioma. A pathology examination was carried out and the results are consistent with MNTI. Angiography reveals a hypervascularized mass in the left hard palate, supplied by branches of the left maxillary artery. Superselective embolization to the feeding artery was done using gelfoam slurry and polyvinyl alcohol (PVA) combination. Two days after embolization, surgical excision was performed with no severe bleeding occurred. No recurrence and surgical complications were found at the 6-month follow-up.

Conclusion: We reported a rare case of hypervascular MNTI in the maxillary region. In a small number of cases, MNTI could exhibit clinical and radiological findings that mimic hemangioma, and histological confirmation is still required for a positive diagnosis. Superselective angiographic embolization is feasible and effective in reducing the risk of severe bleeding during surgical intervention.

EP04

NAVIGATING COMPLEXITY SUCCESSFUL PRE-OPERATIVE EMBOLIZATION OF CAROTID BODY TUMOR ENHANCING SURGICAL PRECISION AND PATIENT OUTCOMES

M. A. Raiman[1], M. A. Kamarudin[1]
[1]Department of Radiology, Hospital Kuala Lumpur, Jalan Pahang, 50586 Kuala Lumpur, Malaysia

Introduction: Carotid body tumor or paraganglioma is one of the rare neoplasms that occur in the head and neck region. Bilateral presentation is even more infrequent, seen in 5% of the population, and is commonly associated with genetic predisposition.

Report: We report a case of bilateral carotid body tumors in a 38-year-old lady, who presented with painless right neck swelling. Imaging findings show features of bilateral carotid body paragangliomas, in which she underwent pre-operative embolization of the right carotid body tumor with subsequent surgical resection. Histopathological examination validated the diagnosis, with immunohistochemistry was positive for SDHD heterozygous pathogenic variant.

Conclusion: Carotid body tumor is a rare disease that may occur at the head and neck region. Although rare, it has characteristic imaging findings that may point out to this diagnosis. Surgery remains as the mainstay of treatment; however, pre-operative tumor embolization is shown to be a safe and efficacious modality that offers reduced peri-operative morbidity. Its applicability should be determined through thorough medical evaluation and discussions between the patient, interventional radiologist, and the surgeon.

EP05

COIL FRACTURE AND UNRAVELLING: AN UNEXPECTED COMPLICATION

W. L. Wong[1], N. Ehsan[1]
[1]Department of Radiology, Hospital Sultanah Aminah Johor Bahru, 80100 Johor Bahru, Malaysia

Introduction: Coil fracture and unravelling are rare complications of coil embolisation of intracranial aneurysms. There has only been a handful of cases found in literature reviews. Hence we describe a case we encountered as well as the management and outcome of this incident.

Results: We report a case of coil fracture and unraveling during embolisation of an aneurysm at the M1/M2 segment of left middle cerebral artery. During the final stage of stent-assisted coil embolisation, there was spontaneous detachment of the coil from its guide wire which was then successfully retrieved. Retrieval was done twice as there was unravelling of the coil fibres, with coil still seen outside of the aneurysm sac raising the concern of partial dislodgement of previously packed coil. Post-retrieval angiogram showed similar appearance of the aneurysm sac as well as the coils within, indicating that the two retrieval attempts were for the same coil which had fractured endovascularly.

Conclusion: Unexpected and rare complications such as endovascular coil fracture can be potentially catastrophic and detrimental to the patient. Hence, awareness of this complication with the skills to promptly manage the situation-in-hand is vital.

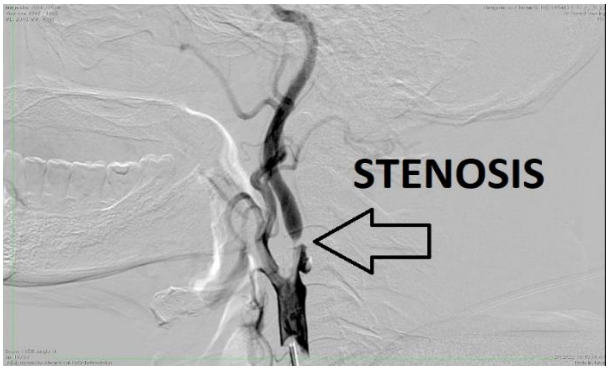
EP06

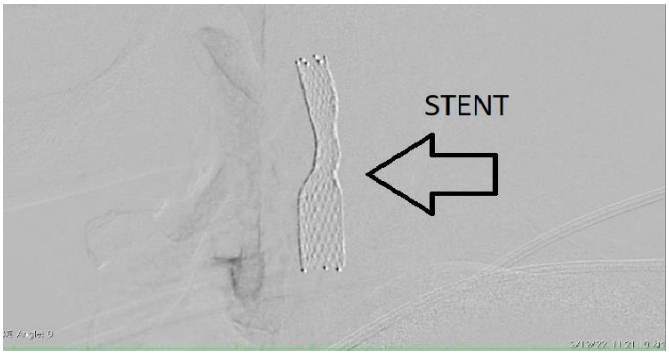
CAROTID ARTERY STENTING (CAS) IN CAROTID ARTERY STENOSIS

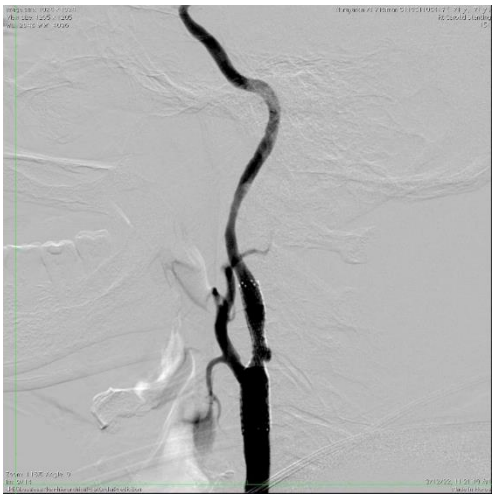
A. Naidu[1], M. A. Kamarudin[1]
[1]Department of Radiology, Hospital Kuala Lumpur, Jalan Pahang, 50586 Kuala Lumpur, Malaysia

Introduction: Carotid artery stenting (CAS). Rather than removing the material in the carotid artery, as is done with CEA, the other alternative is to insert a metal stent (see section on stents) into the artery to push the material out of the way, creating a larger channel for the blood in which clots are less likely to form. There are a number of stents made specifically for the carotid artery that can be used as an alternative to CEA. In addition, stent treatment can be used in other areas of the carotid artery, from the lowest part deep in the chest up to the base of the brain.

Case report: Our patient is a 71-year-old gentleman with previous history of CVA in 2017 with a recent left parietal stroke in November 2022 with right critical carotid artery stenosis. He is also a high-risk patient in view of his underlying heart failure with reduced EF- LVEF 30%. Where he underwent a successful carotid artery stenting via a transfemoral approach. Patient in supine position. Left groin cleaned and draped. Left femoral artery access under US guidance with LA and cannulated using puncture needle. 6Fr short sheath inserted. Angiogram of bilateral CCA and bilateral subclavian artery performed using JB2 and Vert catheter. Initial angiogram showed total stenosis of the left ICA. Both vertebral arteries origin were occluded however there is reconstitution of the vessels from the collateral, also severe stenosis of the proximal right ICA (About 90%) at the level of L3. The short sheath was exchanged with 6Fr long sheath with the aid of exchange glidewire parked in IMA. The long sheath parked at the distal CCA. The stenotic part of the right ICA was crossed using 0.014" wire and Spider Fx was deployed at the distal cervical ICA. The stenosis was pre dilated with Invatec 4x40 balloon. Angiogram performed. Protégé Rx 8-6x40mm Stent was deployed covering the stenotic ICA. Post stenting angiogram performed, Spider Fx was resheathed and removed. Global CCA angiogram performed which showed significant improvement of caliber of proximal right ICA and intracranial vessel caliber. Hemostasis secured by manual compression. Post procedure patient had no immediate complications and was transferred back to the ward uneventfully.







Subsequently after 5 days of observation in the ward which was relatively uneventful patient was fit for transfer back to IJN for continuation of treatment.

Discussion: Carotid artery stenting (CAS) is a minimally invasive technique for treating carotid artery stenosis. Carotid endarterectomy (CEA) has been shown to reduce the incidence of stroke in patients with symptomatic and asymptomatic carotid stenosis [1,2]. However, if the patient is asymptomatic then Offering routine carotid endarterectomy (CEA) or carotid artery stenting (CAS) to patients with asymptomatic carotid artery stenosis (ACS) is no longer considered as the optimal management of these patients. Equally suboptimal, however, is the policy of offering only the best medical treatment (BMT) [5] to all patients with ACS. It is usually indicated in cases such as the above where the patient is unable to tolerate general anesthesia for CEA such as our case who has a severe underlying heart failure with reduced EF- LVEF 30% and known to have cardiac arrhythmia with recurrent ventricular tachycardia on temporary transvenous pacing. No contraindications seen in this patient such as contrast allergy, unfavorable anatomy, unstable carotid plaque and unstable aortic arch plaque. Carotid stenting is equivalent to CEA in reducing carotid stenosis without increased risk for major complications of death/stroke. Because of shortened hospitalization and convalescence, CAS challenges CEA as the preferred treatment of symptomatic carotid stenosis if a reduction in costs can be achieved [3]. Among patients with asymptomatic carotid stenosis however, stenting has a significantly higher rate of any peri-procedural stroke and peri-procedural minor stroke than CEA, and similar risk of peri-procedural major stroke, peri-procedural ipsilateral stroke, or MI [4].

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