B-ENT, 2019, **15**, 371-374

Carotid blowout syndrome in patients with nasopharyngeal carcinoma: three cases

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Key-words. Carotid blowout syndrome; nasopharyngeal carcinoma; radiotherapy.

Abstract. Carotid blowout syndrome (CBS) is a rare complication of radiation therapy for nasopharyngeal carcinoma (NPC), for which angiography is the gold standard for diagnosis and treatment. We report three NPC cases treated with irradiation and complicated by bleeding from the internal carotid artery. The first case presented with multiple episodes of limited nosebleeding followed by massive bleeding. Bleeding was initially stopped by internal carotid artery embolisation. A few months later, meningitis related to skull base osteoradionecrosis occurred and the patient died from sceptic shock. The second case was admitted to the hospital with severe repetitive epistaxis and despite several attempts to manage the bleeding, the patient passed away. The third case presented with a massive epistaxis that resolved itself without any treatment. Spontaneous occlusion of the internal carotid artery occurred without any neurological defects. A nasopharyngeal muscle flap was constructed to prevent meningitis. CBS is a rare but life-threatening complication that requires emergency treatment. Re-irradiation and skull base radioosteonecrosis are strong predisposing factors.

Introduction

Rupture of the internal carotid artery (carotid blowout syndrome [CBS]) is a rare complication of radiation therapy for nasopharyngeal cancer (NPC), resulting from the loss of soft tissue protecting the carotid artery system.[1] The incidence of CBS is 2.6% in patients receiving salvage radiation for head and neck cancer.[2] CBS risk factors include surgery, radiation therapy, pharyngocutaneous fistula, exposure to saliva, wound infection, flap necrosis, and tumour recurrence.[3–5] Endovascular treatment provides an effective way to control the bleeding associated with CBS.[6] Here, we report three cases of irradiated NPC complicated by massive bleeding of the internal carotid artery.

Case reports

Case 1

The first case was a 46-year-old patient diagnosed with right ethmoid esthesioneuroblastoma, who was treated with surgery and radiotherapy. Twenty-five years after these treatments, the patient developed chonic sinusitis and skull base osteoradionecrosis, and subsequently had multiple episodes of epistaxis. Sphenopalatine artery ligation was performed, but 1 month later, the patient suffered again from massive nose bleeding. Then embolisation of the right internal carotid artery was performed and achieved to stop the bleeding (Figs. 1-4). After 6 months, the patient presented a good evolution and neurological defects were limited to the left upper limb. In the same year, he presented with three episodes of meningitis related to sinus infection. Ultimately, he died from septic shock from his final bout of meningitis.

Case 2

A 49-year-old man was transferred to our hospital with severe uncontrolled epistaxis. The patient had been treated for NPC with radiotherapy and chemotherapy 2 years prior. A second irradiation was planned to treat basicranial tumour recurrence.

Conflict of Interest: The authors declare that they no conflicts of interest.
Funding: This article did not require or receive any financial support.
Spring Meeting of the Royal Belgian Society for Oto-Rhino-Laryngology, Head and Neck Surgery, 3 mars 2018.

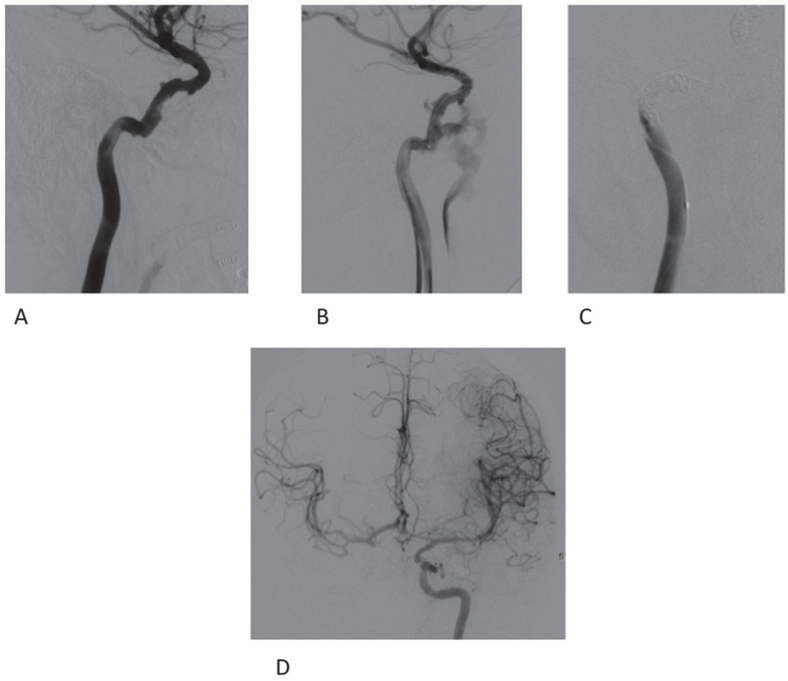

Figure 1

(A) Angiography of Patient #1 showing irregular contours of the right internal carotid artery related to irradiation. (B) Rupture of the right internal carotid artery with massive extravasation of contrast product. (C) Complete occlusion of the internal artery at the end of the embolisation procedure. (D) Selective catheterisation of the left internal carotid artery, which shows a delay of perfusion of the whole right hemisphere.

Bilateral anterior nasal packing was performed in the first hospital, but the physicians were not able to perform posterior nasal packing because of the patient's trimus. Promptly after patient admission to our hospital, he presented with a massive nosebleed that required vesical probe insertion in both nasal cavities and cricothyrotomy to maintain ventilation. Despite these measures, the patient experienced cardiac arrest due to hypovolemic shock. After 15 minutes of cardiopulmonary resuscitation, he was stabilised and angio-CT was performed. Pseudoanevrysm with necrosis of the internal carotid wall was observed as well as severe tumoural stenosis of the internal carotid artery. Thus, no embolisation or surgery was possible and the patient passed away in the intensive care unit.

Case 3

A 44-year-old man presented with one episode of massive epistaxis, which resolved itself without any treatment or compression. His medical history showed that he had been treated for NPC with radiotherapy and chemotherapy 2 decades prior. The treatment was repeated a second time due to early tumour recurrence, but he lived cancer-free for 20 years. Spontaneous occlusion of his internal carotid artery, radio-osteonecrosis of the skull base, and exposure of the dura were found by CT scan and MRI after this bleeding episode (Fig. 2). To prevent meningitis due to this defect, a free anterolateral thigh flap was micro-anastomosed in the nasopharynx at the level of the radioosteonecrosis. This flap initially completed occluded the nasopharynx, and then was debulked 6 months later to restore nasal breathing (Fig. 3).

Discussion

CBS is defined as rupture or exposure of the carotid artery or one of its main branches, associated with bleeding in a patient with a history of head and neck cancer. Irradiation, radical surgery of head and neck tumours, local infection, tumour recurrence, and pharyngocutaneous fistulae are common predisposing factors for this illness.[6] In our case series, the first patient had a right ethmoid esthesioneuroblastoma, presenting with a different

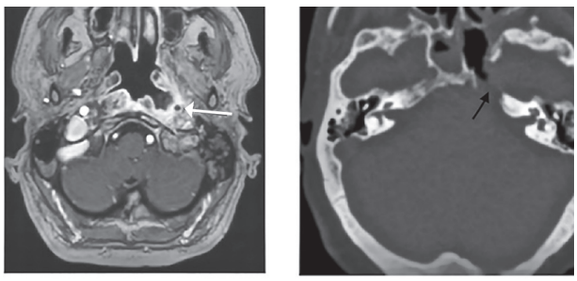

Figure 2

MRI and CT scan of Patient #2 showing osteoradionecrosis of the skull base (black arrow) and complete occlusion of the internal carotid artery (white arrow).

histology and management protocol than the two other patients with NPC. The irradiation protocol differed among the three cases due to the different histologies and management times. The two patients with NPC underwent chemoradiation, whereas the first one patient only underwent irradiation. Associated radiation and chemotherapy, tumour status, and local recurrence may increase the risk of CBS-associated death[7]. However, the risk between patients treated with or without concurrent chemotherapy, or between patients treated with or without salvage surgery is not clearly different according to a previous study.[1]

The mechanism underlying the development of CBS after radiation therapy may be due to radiation-induced vascular injury resulting from obstruction of the vasa vasorum, premature atherosclerosis, advential fibrosis, and necrosis of the arterial wall. Combined with high blood pressure of the great vessel, CBS can result in rupture of the arterial wall and even aortic dissection with extravasation of blood into the pleural cavity.[6,8] Surgery is another iatrogenic factor that may contribute to CBS. Isolated carotid exposure may occur after radical neck dissection in which the sternocleidomastoid muscle is removed. Subsequent dehiscence of the neck incision can then result in exposure and dessication of the vessel wall. This can be avoided by placement of dermal grafts over the vessel or vessel coverage by healthy muscle such as the pectoralis major muscle.[9]

Once repetitive bleeding occurs in a patient with a history of radiation for head and neck cancer, CBS should be suspected and angiography performed as quickly as possible for confirmation. Angiography is the gold standard procedure for CBS diagnosis and offers potential therapeutic management.[6]

CBS can be classified into three clinical entities: threatened, impending, and acute. Threatened CBS is defined as physical or radiologic results suggesting inevitable haemorrhage if no immediate action is taken, such as an exposed artery. Impending CBS is defined as intermittent oronasal bleeding or

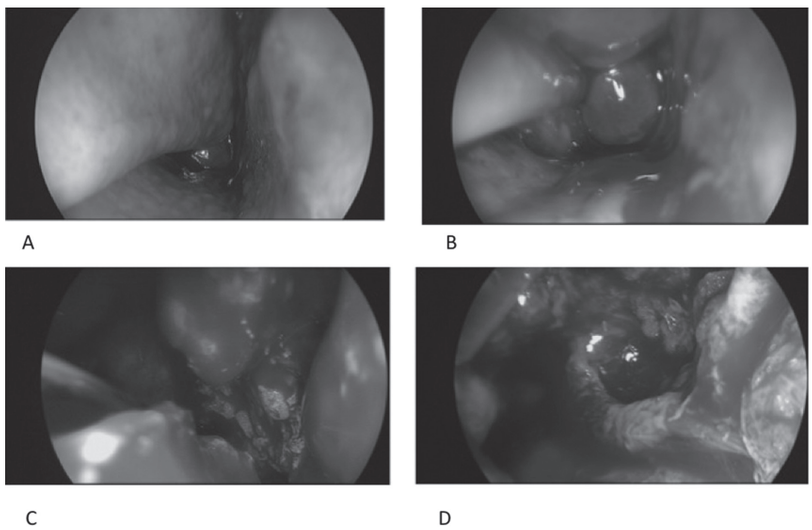

Figure 3

(A) Per-operative pictures of the anterolateral thigh flap debulking procedure in Patient #3, showing entry of the nasal cavity (B) Protruding flap in the cavum. (C) Flap debulking (D) Results at the end of the surgery.

bleeding through a radionecrotic or surgical wound, which can stop spontaneously or by pressure packing. Acute CBS is a profuse haemorrhage that usually cannot be stopped by pressure packing.[1,4,5,6]

Traditionally, emergency surgical ligation has been the only therapeutic treatment available for CBS, with very high mortality and morbidity rates that can reach 40% and 60% respectively. More recently, endovascular treatment has provided an effective way to control the bleeding associated with CBS, by using endovascular coils, balloons, or stents. The best approach for CBS management is to intervene before rupture.[2,6,9]

Conclusion

CBS secondary to radiation therapy is a rare but life-threatening condition, and 76% of CBS cases are fatal. Re-irradiation and skull base radioosteonecrosis are strong predisposing factors. CBS results from the loss of soft tissue protecting the carotid artery system. Early and aggressive management of skull base radionecrosis is indicated to avoid CBS. Once CBS occurs, endovascular treatment is effective to stop the bleeding and is a life-saving intervention.[1,6,9]

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