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Metastatic cerebellar tumor from esophageal squamous carcinoma: a case report and review of the literature

Ludovica Cellini [1], Gonçalo Januário [2], Gonçalo Novais [2]

[1] Policlinico Santa Maria Le Scotte, Siena, Italy

[2] Centro Hospitalar Universitário de Lisboa Central; Hospital de São José, Lisbon, Portugal

Abstract

Esophageal cancer is one of the most fatal malignancies. It is often diagnosed during its advanced stages. Brain metastases, and especially cerebellar ones have been infrequently reported in the Literature. In patients with large cerebellar metastases, surgical removal is still the most effective therapy for improving survival and Karnofsky Performance Score (KFS). Stereotactic Radiosurgery is another treatment option for multiple brain metastases but the improvement in survival was only demonstrated in single metastasis. We report a case of multiple cerebellar metastases from esophageal squamous cell cancer in a 64 years old male, who underwent macroscopic total resection of all lesions. Up to date no treatment guidelines exist for esophageal cancer brain metastases. Multimodal therapy including surgery, radiation, and chemotherapy is now generally recommended for treatment of these lesions. Total resection of cerebellar ones should be the aim of surgical therapy, especially if a good KPS is expected.

Keywords: Brain Metastases; Carcinoma; Craniotomy; Total resection; Radiotherapy; Chemotherapy

Introduction:

Esophageal cancer is one of the most fatal malignancies worldwide, progressively increased in incidence in the Western world. It is the eighth most common incident cancer in the world and it ranks sixth among all cancers in mortality.[1]

The incidence of esophageal carcinoma is approximately 7 cases per 100,000 persons and it has a variable tendency to metastasize to other parts of the body. It is often diagnosed during its advanced stages, because of the lack of early clinical symptoms.[2]

There are two main histological types of esophageal cancer: although esophageal squamous cell carcinoma predominates worldwide, Western nations have seen a marked rise in the incidence of esophageal adenocarcinoma parallel to the increase in obesity.[3]

In 2017 Shaheen et al. performed a review of the literature about esophageal cancer, published on Gastroenterology Research and Practice, analyzing 10049 articles: incidence shows a strong male predominance (84%) with a mean age of 60.7 years. 65% of Esophageal Cancer were located in the lower third of the esophagus .

The two major histological types were adenocarcinoma (40%) and squamous cell carcinoma (60%). The most common sites of esophageal cancer metastases are the lymph nodes, lung, liver, bones and adrenal glands.[4]

The mean incidence of cerebral metastasis ranges from 20% to 40% of patients with solid cancers. Cerebellar and infratentorial metastases are poorly tolerated and carry a grim prognosis compared to supratentorial metastases. Clinical symptoms are often multiple at presentation, most commonly caused by increased intracranial pressure, including headaches (64%), gait disturbances (42%), nausea/vomiting (30%), dizziness (18%), visual changes (10%), altered mental status (6%), and seizure (2%).[5]

At the time of diagnosis, more than 50% of patients present with multiple lesions. In the retrospective study published on Acta Neurochirurgica by Gabriele Schackert et al. in March 2013, they included 127 patients with multiple brain metastases in the study. At the time of diagnosis, more than 50% of the patients already carry multiple lesions, with a mean number of three. The indications for surgery were: large tumours, metastases of unknown primaries at

the time of diagnosis, and space-occupying cerebellar lesions. Gross Total Resection of all simultaneous lesions was necessary to achieve a prolonged survival (limited to 4 lesions in the report). The resection of large lesions, while leaving smaller residual ones, did not result in increased survival.[6]

Brain metastases, and especially cerebellum metastases from esophageal cancer, have been infrequently reported in the literature as a few small series and case reports. The largest series, from Japan, reports the results of 36 patients.[7] Gabrielsen et al. stated the incidence of brain metastasis among esophageal cancer patients as 3.6%. Weinberg et al. reported 27 patients (1.7%) with brain metastasis in a group of 1588 patients with esophageal cancer. Ogawa et al. reported 36 patients (1.4%) with brain metastasis in a series of 2554 patients with esophageal cancer.[7] The prognosis after diagnosis of esophageal cancer brain metastasis is very poor. Once brain metastases are detected, patients generally live for no more than 4 months.[7]

Surgical excision with gross total removal is still the most effective therapy in improving survival and KPS in patients with large cerebellar metastases.[8]

In patients with 2 or 3 brain metastases, with an high performance status and controlled systemic disease, complete surgical resection has good results comparable to those obtained in single lesions.

The Stereotactic Radiosurgery (SRS) is also a good option to treat brain metastases. Based on the Eano Guidelines on this topic, published on Neuroncology by Soffietti et.al in 2017, a single dose SRS in the treatment of a limited number (1–3) of newly diagnosed brain metastases has yielded a local control (shrinkage or arrest of growth) at 1 year of 80%–90% with symptoms improvement and median survival of 6–12 months. Patients with a single lesion, controlled extracranial disease, and KPS of 70% or greater, have longer survival. But in the end the survival advantage was only demonstrated in patients with single metastasis (6.5 vs 4.9 months).[9]

Case report:

We report a case of a male of 64 years old, presented at the ER for headache, dizziness and vomiting progressively worsening in two months. Ineffective therapy with Betahistina Dihydrochloride.

Neurological examination revealed oriented and collaborative patient. Isochoric and isocyclic pupils, without ophtalmoparesis. No nystagmus. Bilateral dysmetria to the nose-index test, especially on the right. Running instability. Absence of focal motor deficits.

CT scan was performed and revealed heterogeneous intra-parenchymal right cerebellar expansive lesion, involving right middle cerebellar peduncle. After contrast, irregular capsula was detected. The CT scan also revealed perilesional edema and signals of hydrocefalus supra-tentorial.

The patient was subjected to a MRI with Gadolinium that revealed three lesions, the major as seen in the CT scan. Other two cerebellar sub-cortical smaller lesions: one on the left side and one in the right side. Below we show the axial (Figure 1), the coronal (Figure 2) and the sagittal (Figure 3) cerebral-MRI scans.

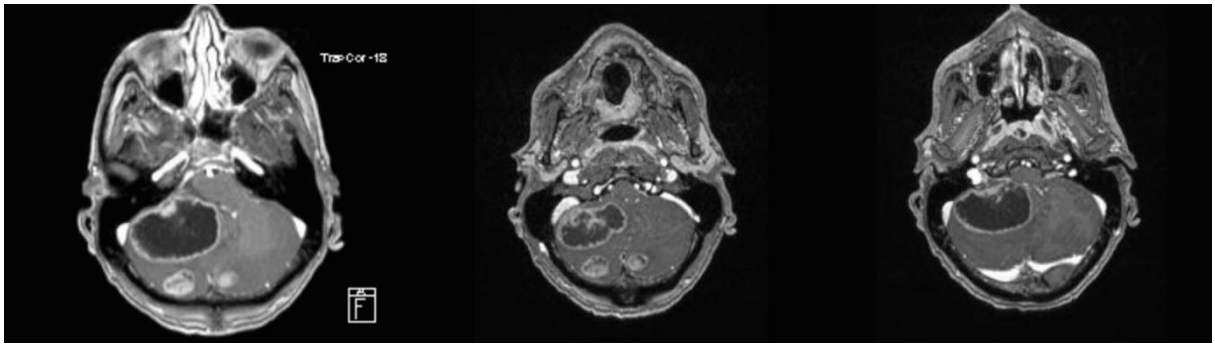

Figure 1 - Axial Cerebral MRI with evidence of three cerebellar lesions. The biggest one on the right side (we can appreciate the contrast-enhancement of the capsula and the cystic/necrotic core). The other two smaller lesions are one on the right side also, and the other, paramedian, on the left side.

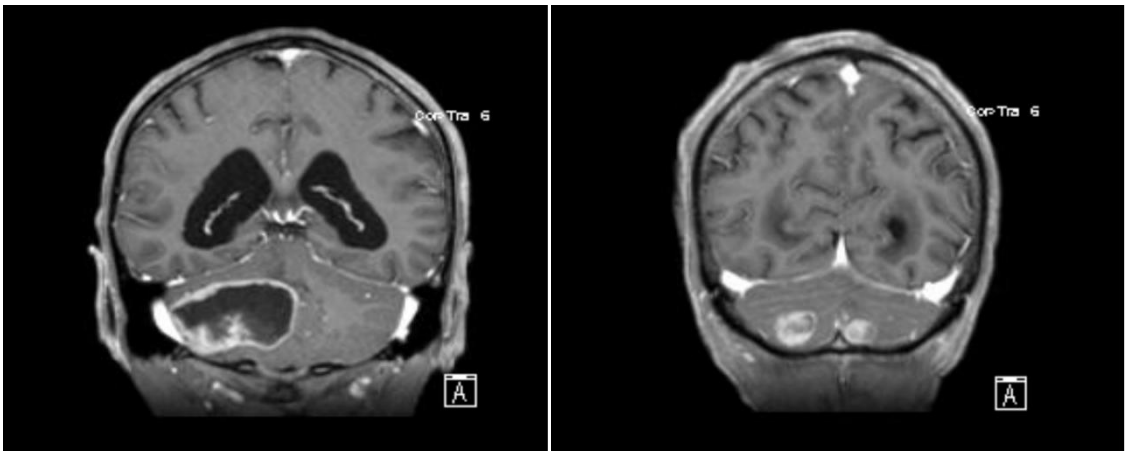

Figure 2 - Coronal Cerebral MRI. The left one documents the largest lesion. The right one the other two smaller lesions.

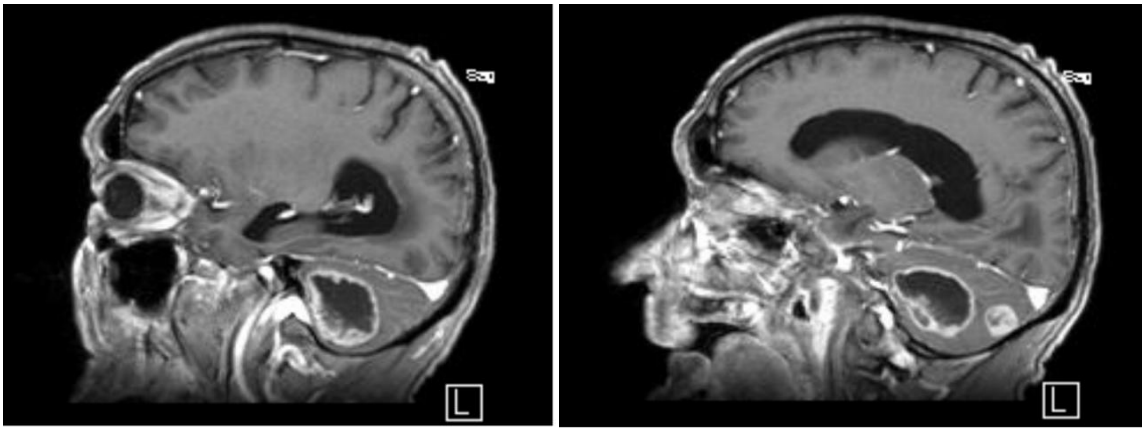

Figure 3 - Sagittal Cerebral MRI revealed the disposition of the lesions in the posterior cranial fossa, helping to decide the most useful approach to remove them.

Total-body CT scan with contrast was performed to stage the patient. It showed atypia of the distal third of the esophagus with periceliac lymph node agglomeration and apparent invasion of the liver capsul. Because of the absence of comorbidities of the patient and the pattern of the localization of the cerebral metastases (that were < 4) the patient was admitted to the Neurosurgical department for open micro-neurosurgical treatment.

The patient was positioned prone, head fixed by a Mayfield headboard in a neutral position. Median incision and sub-occipital craniotomy, mostly lateralized to the right, was performed. We continued with Y-shaped opening of the dura mater and suspension of the flaps. We practiced macroscopically total exeresis of the three lesions with Microscope Leica DHX. Accurate hemostasis was performed and the surgery was without complications.

After the surgery the patient went to the ICU. Patient awakened without complications in the first 24 hours after surgery. Upon awakening, no new onset deficits were detected on neurological examination. We did a post-op CT-scan that revealed gross-total resection of the lesion and the progressive reduction of the ventricular size, without other complications occured. (Figure 4)

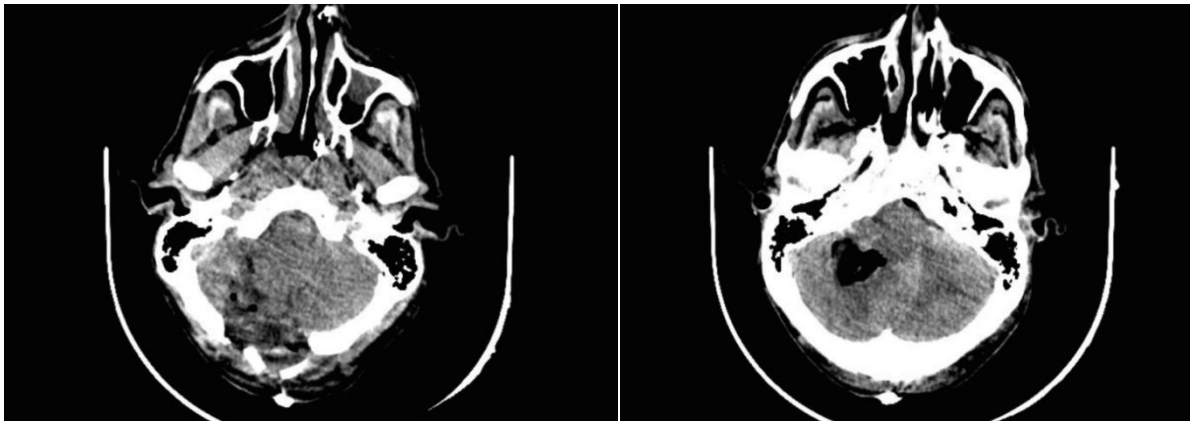

Figure 4 - Axial cerebral CT scan documented the good result of the surgery without complications.

Neurological examination of the patient 3 days after surgery revealed absence of addicional deficit from the pre-op. The histopathological report revealed: cerebellar metastases from primitive esophageal squamous cell carcinoma.

The histopathological report revealed: cerebellar metastases from primitive esophageal squamous cell carcinoma. Immunohistochemical examination showed positivity to Citocheratina CAM5.2, Citocheratina 5/6, P63. Citocheratina 7, 20 and CD56 negative. (Figure 5)

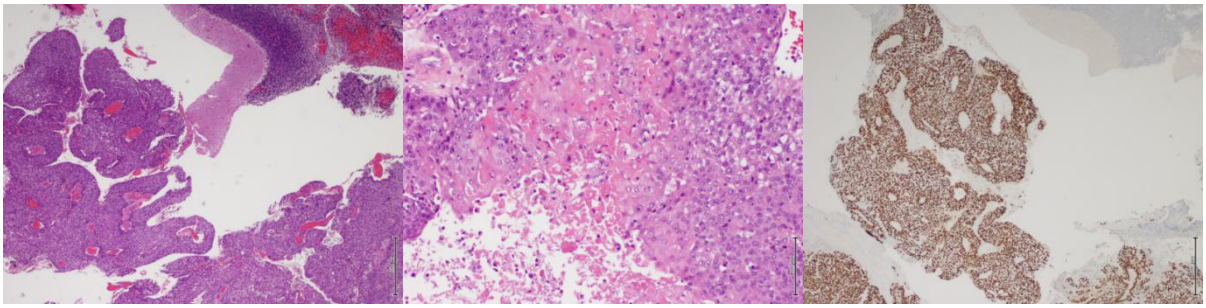

Figure 5: Histological slides of the lesions showed the typical pattern of esophageal squamous cell carcinoma characterized microscopically by keratinocyte-like cells with intercellular bridges or keratinization.

Discussion:

Esophageal cancer is a rare nosological entity, progressively increasing, especially in Western countries, due to eating habits. The diagnosis is often late, due to the absence of the symptoms of the primary tumor. Once diagnosed, the tumor frequently presents itself in stage IV. The treatment of esophageal carcinoma depends on the stage of presentation.

Brain metastases from esophageal cancer are rare. Cerebellar metastases are even rarer and also the most clinically symptomatic ones. Multiple brain metastases, as evidenced in some studies, have a favorable outcome after complete resection (in cases of 1 to 3 intracranial metastases).

Our report concerns a rare case of a patient with three cerebellar metastases from esophageal cancer, treated with microneurosurgery. We have decided infact, given the age, the good KPS of the patient and the location of the cerebellar metastases, to remove all three lesions, using the same approach (sub-occipital craniotomy).

As recommended for typical brain metastases, we performed gross total resection and adjuvant WBRT at the end of the first month after surgery. There were no complications during the surgical procedure and the patient, awakened during the first post-operative day, showed a neurological examination without worsening compared to pre-operative. We will follow the patient clinically and radiologically, subjecting him to a control MRI within 3 months.

Conclusion:

Due to the lesion's rarity, there are no treatment guidelines for these patients. Multimodal therapy including surgery, radiation, and chemotherapy is now generally recommended for treatment of brain metastases of esophageal cancer.

Surgical resection should be considered in patients with a limited number (1 to 3) of newly diagnosed brain metastases, especially in case of lesions of ≥3 cm in diameter (symptomatic or not), lesions with necrotic or cystic appearance and edema/mass effect, lesions located in the posterior fossa with associated hydrocephalus, and lesions located in symptomatic eloquent areas.[9]

The Sterotactic Radio-surgery also represents nowadays a good choice to treat multiple brain metastases and should be considered as first line of treatment in patients with metastases that are not resectable due to location or with comorbidities precluding surgery.[9]

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