CASE REPORT

Neuroembolization of metastatic Merkel cell cancer to the face for treatment of Kasabach–Merritt syndrome

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SUMMARY

Kasabach–Merritt syndrome is defined as a consumptive thrombocytopenia in the presence of a highly vascular tumor. Multiple treatment options, including transarterial embolization, have been described. We demonstrate that transarterial embolization is a viable option in the treatment of a rapidly progressive and debilitating Merkel cell tumor metastasizing to the head and neck presenting with Kasabach–Merritt syndrome.

BACKGROUND

Kasabach–Merritt syndrome is defined as a consumptive thrombocytopenia in the presence of a highly vascular tumor. Multiple treatment options have been described in this setting.¹ Stanley *et al*² first described the treatment of Kasabach–Merritt syndrome secondary to a buttock hemangioma in an infant using transarterial embolization (TAE) with polyvinyl alcohol (PVA). Since then, there have been reports of Kasabach–Merritt syndrome presenting with head and neck hemangiomas being treated with TAE.³ We demonstrate that TAE is a viable option in the treatment of a rapidly progressive and debilitating metastatic tumor to the head and neck presenting with Kasabach–Merritt syndrome.

CASE PRESENTATION

A 69-year-old woman with a past medical history of Merkel cell cancer metastatic to the right cheek was admitted for regional relapse of tumor extending over her right face (figure 1). She had previously been treated over the past year with multiple surgeries and multiple courses of chemotherapy and radiation therapy. Systemic agents included carboplatin, etoposide, temazolamide, hycamtin, and doxorubicin. Surgery included resection of multiple skin lesions, including a third operation 8 weeks prior to admission. The lesions had subsequently grown back in a short time. The patient was noted to have coagulopathy and thrombocytopenia, which were presumed to be secondary to Kasabach–Merritt syndrome. The pretreatment platelet count was 23 000/ μ l and was associated with purpura, ecchymosis, and mucosal bleeding.

INVESTIGATIONS

Imaging by CT showed an extensive exophytic, hypervascular soft tissue mass involving the right temporal, facial, premaxillary, superior and inferior palpebral, auricular, and preseptal regions (figure 2). There was no associated bony destruction. The

tumor supply was predominately from the right facial artery and to a lesser degree the right superficial temporal, anterior and middle deep temporal, posterior auricular, and internal maxillary arteries.

TREATMENT

Given the rapid increase in size of the tumor despite multiple resections and chemotherapy/radiation therapy and the presence of Kasabach–Merritt syndrome, vascular embolization was discussed with the patient.

After the standard sterile preparation, the right external carotid artery was subselected, demonstrating a markedly hypervascular tumor involving the

Figure 1 Frontal photograph of the patient, demonstrating bulky metastatic lesions extending over the right face.

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Figure 2 Coronal post-contrast CT image demonstrating multiple hypervascular lesions overlying the patient's right face.

right subcutaneous face. Multiple feeder external carotid artery branches were identified, including the distal internal maxillary sphenopalatine branches, anterior deep temporal branch, facial artery, occipital artery, and posterior auricular artery (figure 3).

Using a roadmap technique, each branch (except for the right occipital artery) was selected coaxially using a Renegade microcatheter and Synchro wire through a Berenstein catheter. Each branch showed a hypervascular supply to the tumor and was embolized using 150–350 μ m PVA particles (figure 4). The right occipital artery was coaxially subselected using an SL 10

Figure 3 Selective external carotid angiography demonstrates prominent tumor blush from multiple feeder vessels.

Figure 4 Selective external carotid angiography after embolization with polyvinyl alcohol particles demonstrating significant tumor devascularization.

microcatheter and Synchro wire. Hypervascular supply to the tumor was noted and 6 mg of methohexital (Brevital Sodium) were injected into the vessel so that neurological testing could be performed. The patient did not experience any neurological changes and the tumor was embolized using 50–150 μ m PVA particles. Follow-up angiogram of the right common carotid artery demonstrated approximately 70% decrease in tumor devascularization. Persistent supply to the tumor from the right ophthalmic artery and to a lesser degree from a proximal branch of the right occipital artery was noted (figure 5).

Figure 5 Selective angiogram of the internal carotid artery demonstrates persistent vascular supply predominately via the ophthalmic artery and to a lesser degree a proximal branch of the occipital artery.

OUTCOME AND FOLLOW-UP

Follow-up CT scan demonstrated a significant decrease in size and attenuation of the right cheek mass (figure 6). The central low density of the mass was thought to represent post-embolic oligemia. Physical examination also demonstrated significant improvement in size of the tumor (figure 7). The post-treatment peak platelet count was 76 000/ μ l and was noted to be 42 000/ μ l on discharge 1 week post-procedure. The platelet count subsequently returned to near baseline levels of 108 000 4 weeks after the procedure with resolution of bleeding.

DISCUSSION

Initially, TAE was primarily used in head and neck cancer for the treatment of intractable hemorrhage.⁴ TAE has since been part of combined efforts at treatment of two primarily head and neck cancers: paragangliomas and juvenile nasopharyngeal angiofibroma.^{5 6}

Kasabach–Merritt syndrome is defined as consumptive thrombocytopenia as a result of a highly vascular tumor. Multiple treatment options have been described in the treatment of Kasabach–Merritt syndrome with multimodality treatment being favored more recently.¹ Stanley *et al*² first described the treatment of Kasabach–Merritt syndrome with TAE using PVA in a buttock hemangioma. Their treatment resulted in both cosmetic improvement and resolution of the coagulopathy. Since then, others have reported treatment of Kasabach–Merritt syndrome secondary to head and neck hemangiomas with TAE.³ Our patient's Kasabach–Merritt syndrome manifested as a result of highly vascular metastatic Merkel cell cancer metastatic to the right cheek region. The recurrence of tumor despite multiple surgeries and cycles of chemotherapy and radiation

Figure 6 Coronal post-contrast CT image demonstrating significant decrease in the size of the right face tumor masses.

Figure 7 Frontal photograph of the patient demonstrating significant improvement in the size of the right face tumor masses.

therapy presented TAE as the main viable treatment strategy. As previously reported, our patient's platelet counts also improved dramatically after TAE of the tumor.

Head and neck cancer is very debilitating, in large part due to physical symptoms such as dysphagia, pain, and speech problems, and psychosocial issues such as fatigue and depression.⁷ Our patient complained of similar issues on her recent admission. On follow-up, the patient reported improvement in her symptoms and was also noted to have a cosmetic improvement given the decrease in tumor bulk.

Learning points

- ▶ Transarterial embolization is a viable option for treatment of Kasabach–Merritt syndrome in the setting of hypervascular metastatic disease to the head and neck.
- ▶ Improvement in consumptive thrombocytopenia is an expected result after treatment.
- ▶ Improvement in physical symptoms and psychosocial issues can also be seen due to decrease in tumor size.

Contributors All authors participated fully in the conception and design of this case report and were actively involved in the drafting of the manuscript and in the final submission process.

Competing interests None.

Patient consent Obtained.

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