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CASE REPORT

BEGINNER

HEART CARE TEAM/MULTIDISCIPLINARY TEAM LIVE: SURGERY AND INTERVENTIONS

Type A Aortic Dissection With Concurrent Aortic Valve Endocarditis, Subarachnoid Hemorrhage, and Disseminated Intravascular Coagulation

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ABSTRACT

We describe surgical repair of a Stanford Type A aortic dissection with concurrent aortic valve *Streptococcus equi* endocarditis in the setting of subarachnoid hemorrhage and disseminated intravascular coagulation. Multidisciplinary collaboration among specialists from a variety of disciplines is essential when treating acutely ill cardiovascular patients with multisystem involvement. (**Level of Difficulty: Beginner.**) (J Am Coll Cardiol Case Rep 2022;4:839–843)

CLINICAL SUMMARY

A 57-year-old man with a history of recent dental work was taken to the emergency department after being found at home with an altered mental status, urinary incontinence, and generalized weakness. His family reported that he had 2 weeks of malaise, fever, emesis, and diarrhea before presentation. Computed tomography (CT) of the head revealed an intracranial hemorrhage (**Figure 1**) and dissection flap within the aortic arch (**Figure 2**). Our center was contacted for potential transfer to Stanford Hospital. Subsequent CT angiography of the chest, abdomen, and pelvis confirmed a 6.5-cm Stanford Type A aortic dissection from the root to the external iliac arteries (**Figure 3,** Video 1). He was transferred to Stanford for further care.

LEARNING OBJECTIVES

- To recognize that surgical repair of an acute Type A aortic dissection should always be considered, even in the setting of multiple relative contraindications or risk factors.
- To identify the potential clinical manifestations of a rare but often fatal bacterial pathogen, *S. equi*.
- To advocate for collaboration between multidisciplinary teams, which is essential when caring for complex patients with involvement of multiple organ systems
- To demonstrate the value of preoperative echocardiography in surgical planning, when time allows.

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ABBREVIATIONS AND ACRONYMS

CT = computed tomography

GCS = group C streptococci

SAH = subarachnoid hemorrhage

His vitals on arrival were temperature 39.1°C, heart rate 106 beats/min, blood pressure 67/32 mm Hg, and SaO_2 97%. The patient was alert and oriented but slow to respond, with right upper and lower extremity weakness. Laboratory data are shown in the **Table 1**.

WHAT ARE THE NEXT STEPS IN THE WORK-UP OF THIS PATIENT?

Type A aortic dissection is a surgical emergency associated with a 1%-2% increase in mortality for every hour that surgery is delayed.[1] The initial head CT showed a small subarachnoid hemorrhage (SAH) (**Figure 1**). Neurocritical care and neurosurgery were consulted to assist in risk stratification before possible operative repair and systemic heparinization during cardiopulmonary bypass. Bedside transthoracic echocardiography demonstrated a mobile mass in the left ventricular outflow tract (Video 1). The patient was started on empiric vancomycin and piperacillin-tazobactam after blood cultures were sent. Gram stain was notable for Gram-positive cocci. The patient met both major Duke Criteria and 1 minor criteria (vascular phenomena) for infective endocarditis.

FIGURE 1 Computed Tomography of the Head Without Contrast

Computed tomography of the head without contrast showed a subarachnoid hemorrhage in the right parietal lobe. The hemorrhage is indicated by the subtle hyperdensity within the **red ellipse**.

HOW DOES THE PATIENT'S SAH AFFECT HIS PERIOPERATIVE MANAGEMENT?

After an extensive discussion with the neurocritical care team and the patient, a collective decision was made that the risks of delaying surgery outweighed the risk of systemic heparinization and cardiopulmonary bypass. Per neurosurgery, there was no indication for neurosurgical intervention, because the patient had no midline shift or evidence of elevated intracranial pressure. Neurocritical care recommended attempting to normalize the international normalized ratio (INR) to <1.5 and platelets to 75,000-100,000, and to perform intraoperative neurological pupil index and postoperative imaging of the head.

WHAT COULD EXPLAIN THE PATIENT'S HEMATOLOGICAL ABNORMALITIES AND HOW SHOULD THEY BE CORRECTED IN THE PERIOPERATIVE PERIOD?

Although aortic dissections can cause consumptive coagulopathy and thrombocytopenia, the cause of this patient's thrombocytopenia is likely disseminated intravascular coagulation secondary to sepsis. The patient received 2 U of fresh frozen plasma (FFP) and 3 U of platelets preoperatively, with an improvement of his INR to 1.5 and platelets to 103,000. He received 4 U of packed red blood cells, 4 U of cryoprecipitate, 6 U of fresh frozen plasma, and 7 U of platelets during the procedure. His coagulopathy was inevitably exacerbated by systemic heparin during cardiopulmonary bypass, and he received 3 vials of activated prothrombin complex concentrate at the end of the procedure. On arrival to the cardiovascular intensive care unit, his INR had improved to 1.2, partial thromboplastin time 30.6, and platelets of 125,000.

HOW WOULD ENDOCARDITIS OF THE AORTIC VALVE WITH CONCOMITANT TYPE A DISSECTION AFFECT MANAGEMENT?

The 2019 American Association for Thoracic Surgery Guidelines on Surgical Management of Infective Endocarditis[2] give surgery a Class I, Level of Evidence: B recommendation for patients with valve dysfunction and heart failure symptoms. Transthoracic echocardiography showed Type A dissection with moderate-severe aortic valve insufficiency secondary to a mobile mass in the left ventricular outflow tract (Video 1), and an ejection fraction of 47%. The 2010 American Heart Association guidelines on the management of acute ascending aortic

FIGURE 2 Computed Tomography Angiography of the Head and Neck

Computed tomography angiography of the head and neck demonstrated a dissection flap in the aortic arch, indicated by the **red arrows**, which is concerning for Type A aortic dissection.

dissections recommend urgent evaluation for surgical repair (Class I, Level of Evidence: B).[3] In the context of the patient's multiple indications for emergent surgery, the decision to proceed with operative management is supported by current guidelines.

WHAT OPERATION SHOULD THIS PATIENT UNDERGO?

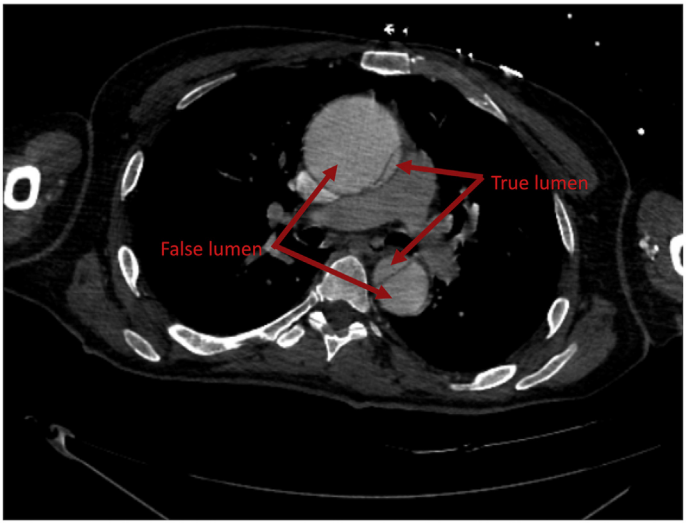
The patient requires Type A aortic dissection repair and an aortic valve replacement given the size of the vegetation and destruction of the valve leaflets. He underwent complete replacement of his aortic root with a bioprosthetic aortic valve within a composite-valve-graft and hemi-arch replacement. Intraoperative findings included a dissection flap extending into the aneurysmal root, purulent thrombus within the false lumen (intraoperative cultures sent, discussed in the following text), and replacement of the noncoronary cusp by a large vegetation with partial involvement of the left and right cusps. The descending thoracic aorta was nonaneurysmal at the time of surgery, measuring 3.6 cm. Based on the CT angiography, there was no evidence of malperfusion of the abdominal viscera, which were perfused via the true lumen.

BLOOD CULTURES AND SURGICAL SPECIMENS FROM THE AORTIC VALVE VEGETATION AND EXCISED AORTA GREW *STREPTOCOCCUS EQUI*, GROUP C STREPTOCOCCI. WHAT IS KNOWN ABOUT THIS PATHOGEN?

The pathogen *Streptococcus equi (S. equi)* is typically considered an opportunistic infection in humans, but can be highly virulent with reported mortality as high as 25% depending on the site of infection and patient age.[4] *S. equi* rarely causes bacteremia, and even less frequently causes endocarditis. In 1 study of over 150,000 blood cultures, only 8 grew *S. equi* and only 1 was diagnosed with endocarditis.[5] There are several reports of *S. equi* endocarditis in Europe.[6–8] Notably, most patients who contract group C streptococci (GCS) bacteremia have an underlying malignancy, cardiovascular disease, recent consumption of unpasteurized dairy products, or contact with horses.[4] One report described canine to human transmission of GCS; however, the canine in the case report developed respiratory symptoms.[9] Our patient confirmed walking his dog on trails also used for equestrian activities, but denied any illness in his dog. He denied risk factors for both development of Type A dissection and GCS bacteremia, other than a right maxillary bone graft several months prior. Subspeciation of *S. equi* was not available, and has proven difficult for many laboratories caused by frequent misidentification.[8]

Our infectious disease specialists were consulted to advise on the treatment of this unusual pathogen with multisystem involvement. He was initially treated with empiric vancomycin and piperacillin-tazobactam. Once blood and intraoperative cultures grew *S. equi*, he was narrowed to intravenous penicillin for a 6-week course. On postoperative day (POD) 5, he developed transient fevers without leukocytosis and underwent an extensive work-up for disseminated infection. Magnetic resonance imaging during the postoperative infectious work-up showed punctate infarcts with associated microhemorrhages in the brain, as well as evidence of osteomyelitis or discitis at the level of C4-C5, for which he was already on the

FIGURE 3 Computed Tomography Angiography of the Chest, Abdomen, and Pelvis



Computed tomography angiography of the chest confirmed the suspected Type A dissection. The true and false lumen are labeled accordingly.

TABLE 1 Admission Laboratory Data

	Value	Reference Range
WBC, K/uL	13.8	4.0-11.0
Hgb, g/dL	11.7	13.5-17.7
Hct, %	33.8	40-52
Platelet count, K/uL	28	150-400
MCV, fL	94.4	82.0-98.0
RDW, %	13.1	11.5-14.5
RBC, MIL/uL	3.58	4.4-5.9
MCH, pg	32.7	27.0-34.0
MCHC, g/dL	34.6	32.0-32.6
Na, mmol/L	139	135-145
K, mmol/L	3.5	3.5-5.5
Cl, mmol/L	106	98-107
CO_2, mmol/L	23	22-29
Anion gap, mmol/L	10	5-15
Glucose, mg/dL	135	70-140
BUN, mg/dL	42	6-20
Cr, mg/dL	1.25	0.67-1.17
Ca, mg/dL	7.7	8.4-10.2
Mg, mg/dL	2.8	1.6-2.6
D-dimer, ug/mL FEU	5.3	<0.50
Fibrinogen, mg/dL	267	234-395
PT, s	23.5	11.5-14.7
PTT	32.6	23.8-35.7
INR	2.2	0.9-1.2

BUN = blood urea nitrogen; Ca = calcium; Cl = chloride; Cr = creatinine; FEU = fibrinogen equivalent units; Hct = hematocrit; HgB = hemoglobin; INR = International Normalized Ratio; K = potassium; MCH = mean corpuscular hemoglobin; MCHC = mean corpuscular hemoglobin concentration; MCV = mean corpuscular volume; Mg = magnesium; MIL = million; Na = sodium; PT = prothrombin time; PTT = partial thromboplastin time; RBC = red blood cell; RDW = red cell distribution width; WBC = white blood cell.

appropriate antibiotic regimen, penicillin. The patient was also evaluated by ophthalmology for vitreitis, which was treated empirically with intravitreal vancomycin and ceftazidime, although it was later believed to be more consistent with ocular ischemic syndrome secondary to Type A dissection. Dentistry was consulted and he was found to have severely decayed dentition and a probable abscess at the site of a prior right maxillary bone graft with an open tract between the right maxillary sinus and oral cavity. He was taken to the operating room for debridement, washout, and repair, and tolerated the procedure well without complications. Following this procedure, he remained afebrile for the remainder of this hospital stay.

Notably, other than the initial blood cultures drawn on admission and the intraoperative cultures, which both grew *S. equi*, all subsequent bacterial cultures were negative during his hospital stay. Upon discharge, a peripherally inserted central catheter was placed for 6 weeks of penicillin G, with the ultimate duration to be determined by trending inflammatory markers as an outpatient, in accordance with the recommendations from our infectious disease colleagues.

REMAINDER OF HOSPITAL COURSE

On POD1, the patient spontaneously moved all extremities and repeat head CT demonstrated the stable SAH. He was extubated on POD2 and transferred from the cardiovascular intensive care unit to the ward on POD8. By POD30, his mentation and dysarthria improved with only mild word-finding difficulty. The patient's mobility improved, platelets normalized (249,000), and he was discharged to a rehabilitation facility on POD30 with 6 weeks of penicillin G. Surgical pathology demonstrated acute inflammation and Gram-positive cocci within the aortic wall. The final pathological diagnosis was mycotic aortic aneurysm with dissection and aortic valve endocarditis.

Four months after the index procedure, the patient returned to the emergency department with pleuritic chest pain and was readmitted for CT findings of contrast extravasation within a perigraft fluid collection surrounding the root and ascending aortic graft. He subsequently underwent redo sternotomy and replacement of his aortic and mitral valves for dehiscence of the aorto-mitral curtain secondary to recurrent endocarditis. He was discharged on POD16 with lifelong amoxicillin-clavulanate, per the infectious disease team.[10] He returned to clinic for his 4-month follow-up appointment and has remained healthy, but is struggling with insurance issues and the cancellation

of physical therapy appointments caused by the COVID-19 pandemic.

CLINICAL PERSPECTIVES

This case demonstrates that surgical repair of a Stanford Type A aortic dissection with concurrent aortic valve endocarditis, thrombocytopenia, and SAH is feasible, which to our knowledge, has not been reported in the published literature prior to this report. This case demonstrates the importance of having a multidisciplinary team of specialists to evaluate and manage complex cases, as well as the necessity for expeditious transfer of similar patients to centers with expansive multidisciplinary resources.

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KEY WORDS critical care, endocarditis, group C streptococcus multidisciplinary, *Streptococcus equi*, type A

APPENDIX For a supplemental video, please see the online version of this paper.