Aortic dissection in a patient with Turner syndrome

Disección de aorta en una paciente con síndrome de Turner

To the Editor,

Turner syndrome is an X-chromosome monosomy (X0). Patients with this syndrome are infertile, have short stature, and, among other abnormalities, may present cardiovascular anomalies in up to 55% of cases. One of these abnormalities is aortic dilatation, present in 3–42% of patients, while aortic dissection is less frequent but potentially fatal.[1,2]

We report the case of a 23-year-old woman from the Philippines with a history of Turner syndrome, arterial hypertension, dyslipidemia, amenorrhea, and polycystic ovary syndrome. Her outpatient treatment included valsartan/hydrochlorothiazide 320 mg/25 mg every 24 hours, atorvastatin 60 mg every 24 hours, and amlodipine 10 mg every 24 hours. She was brought to the emergency department by the Emergency Medical Service after falling backward in a restaurant, following the sudden onset of retrosternal chest pain and a sensation of lightheadedness. Physical examination showed a blood pressure of 132/81 mmHg, heart rate of 70 beats/min, and oxygen saturation of 99%. Systemic examination was normal, but neurologic evaluation revealed bilateral flaccid paralysis with areflexia of both lower extremities. Laboratory tests, electrocardiogram, and cranial computed tomography (CT) were unremarkable. Thoracoabdominal CT revealed a rupture of the intimal layer of the ascending thoracic aorta with formation of a double lumen and retrograde extension into the brachiocephalic trunk, right common carotid artery, left subclavian artery, and probably the ostium of the right coronary artery. Antegrade extension was observed along the abdominal aorta, involving the celiac trunk, superior mesenteric artery, and right renal artery, reaching the infrarenal abdominal aorta. A thrombus was identified extending into both common iliac arteries, internal iliac arteries, and proximal external iliac arteries, with re-permeabilization of the right distal external iliac artery and the left proximal external iliac artery. Distal thrombosis of the superior mesenteric artery and complete thrombosis of the inferior mesenteric artery were also noted, along with hemopericardium and signs of visceral ischemia in the right kidney, as well as concomitant renal, splenic, and hepatic infarctions. The final diagnosis was a Stanford type A aortic dissection with secondary visceral ischemia (Figure 1).

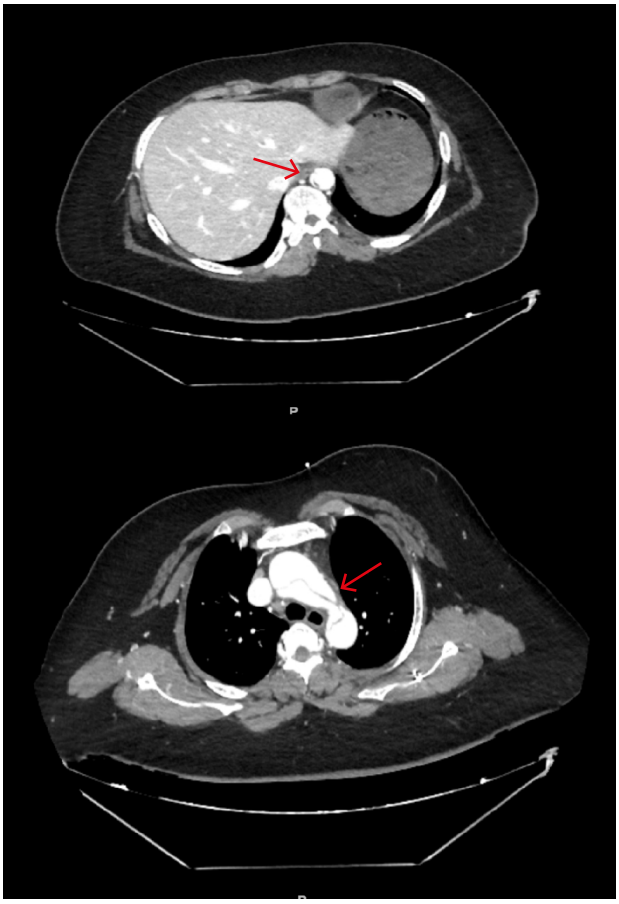

Figure 1. Descending and ascending aorta with double arterial lumen.

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Given these findings, the patient was transferred to a referral center for surgery, where ascending aorta replacement and aortic valve commissural resuspension were performed. During the postoperative period, she developed right-sided colonic ischemia requiring right hemicolectomy. Her clinical course worsened, and she died 48 hours after surgery due to distributive shock.

In Turner syndrome, cardiovascular abnormalities are present in up to 55% of patients. Reported abnormalities include bicuspid aortic valve (15–30%), elongation of the transverse aortic arch (40–50%), coarctation of the aorta (7–18%), and aortic dilatation (3–42%), whose dissection is potentially fatal, although at times asymptomatic.[1,2] Therefore, cardiologic follow-up in these patients is essential. In the case presented here, bilateral flaccid paralysis of the lower extremities was caused by involvement of the anterior segmental medullary artery, known as the artery of Adamkiewicz. Because of its location, this artery is one of the main contributors to the anterior spinal cord blood supply, and its compromise can produce an anterior spinal artery syndrome due to spinal cord ischemia.[3-5] In patients with Turner syndrome presenting with chest pain and associated neurologic symptoms, aortic dissection should be considered in the differential diagnosis because of its potentially fatal consequences.

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Forty years of Emergency Medical Services by the city of Zaragoza's fire department

Cuarenta años del servicio de asistencia médica del cuerpo de bomberos del Ayuntamiento de Zaragoza

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To the Editor,

We have read with interest the special article by Pacheco-Rodríguez, recently published in Revista Española de Urgencias y Emergencias, in which he reviews his experience in the evolution of emergency medicine in Spain.[1] Although he cites some Emergency Medical Services (EMS) belonging to fire departments, we believe that the EMS of the Zaragoza Fire Department (Medical Assistance of the Firefighting, Rescue, and Civil Protection Service of the Zaragoza City Council (Zaragoza, Spain)–AMBZ–), created in 1983, deserves an individual mention on the occasion of its 40th anniversary.[2]

The AMBZ EMS has been a pioneer in out-of-hospital emergency care at the regional level,[3] being the first service to perform prehospital advanced life-support procedures in Aragón and during incidents involving multiple victims (IMV).[4] Nationwide, we also introduced innovative interventions[5] and were the service that provided the first out-of-hospital treatment of cyanide poisoning through the administration of hydroxocobalamin,[6] as well as inhaled loxapine in cases of psychomotor agitation.[7]

This service drafted the Plan for the Organization and Out-of-Hospital Health Response to Municipal Emergencies, approved by the Zaragoza City Council in 1993, which was one of the first plans of its kind in Spain. It was presented at the First International Meeting on Emergency and Disaster Medicine held in Madrid in 1994 and has been updated over the years, the latest version dated April 11th, 2023. Under this municipal plan, AMBZ coordinates and leads the on-scene health group response in IMV events.[8]

The service's operational area is the Municipality of Zaragoza, and it is activated

Table 1. Human and material resources

Human resources	Material resources
1 Medical Chief	3 Advanced Life Support ambulances
13 Physicians	2 Advanced medical posts
13 Nurses	1 Rapid Response Medical Vehicle
36 Firefighter–EMTs	1 Rapid decontamination vehicle
12 EMTs	

EMT: Emergency Medical Technician.


First AMBZ ambulance in (1983).


1985 ambulance.


Ambulance interior (2013).


AMBZ personnel team.

Figure 1. Evolution of the mobile resources of the AMBZ service.
AMBZ: Medical Assistance of the Firefighting, Rescue, and Civil Protection Service of the Zaragoza City Council.

by 112-Aragón, 061-Aragón, the police, or directly by the public. It currently has the staffing and intervention resources summarized in Table 1. With these resources, we provide the city of Zaragoza with one advanced life-support ambulance operating 24/7/365 and a second unit operating 14/7/365. Our annual activity amounts to approximately 3,000 emergency calls. Since the service's inception, 91,963 ambulance interventions have been performed.[9] Our clinical activity is coordinated with the other EMS systems in Zaragoza and Aragón (Spain), and we are included in the regional STEMI and Stroke Codes. Since 2019, the service has had access to the Aragonese Electronic Health Record through tablets carried in the ambulances. Similar to the Unitat d'Intervenció i Suport (UIS) of the SEM in Catalonia (Spain) or Dispositivo Especial Preventivos Actos Antisociales (DEPAS) of SAMUR-PC in Madrid (Spain), the AMBZ provides medical coverage for the Special Operational Security Group of the National Police in Zaragoza during interventions that involve significant risk for its members.[10] Furthermore, just like any other EMS, the service carries out extensive training activities, both internally and for the rest of the Fire Department, medical and nursing students, other emergency professionals, and the general public.[7]

AMBZ continues to engage in important multicenter research, together with other EMS in Spain,[11] including the Spanish Registry of Out-of-Hospital Cardiac Arrest (OHSCAR),[12] the Study on Care Quality and Health Outcomes in Time-Sensitive Conditions Treated by Out-of-Hospital Emergency Services (IMPACTE), and the Study of Out-of-Hospital Emergency Services in the Management of the Severe Trauma Patient (ESTRAGE).

Although we are a small EMS, operating at the municipal level and with far fewer resources than other Spanish services, we believe that our 40 years of activity have allowed us to develop a significant body of clinical, training, and research experience. This has earned us a pioneering place in the historical development of emergency and prehospital care in Spain

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Usefulness of bedside ultrasound in the emergency diagnosis of splenic rupture: a case report

Utilidad de la ecografía clínica a pie de cama en el diagnóstico de la rotura esplénica en urgencias: a propósito de un caso

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To the Editor,

The spleen is an immunologic organ whose rupture is most widely caused by trauma, with atraumatic rupture being less frequent and having a prevalence of 0.1–0.5%.[1]

We present the case of a 58-year-old woman, smoker, who presented with sharp abdominal pain rated 7 on the Visual Analog Scale (VAS), of sudden onset in the left hemiabdomen and radiating to the ipsilateral hemithorax and shoulder, predominating in the supine position. She reported no other symptoms. She described an emetic episode and increased bowel movements during the prior 3 days.

On examination, she had a preserved airway, good bilateral air entry, slight cutaneous pallor, present and symmetric radial pulse, capillary refill < 2 seconds, blood pressure 116/65 mmHg, heart rate 66 bpm, Glasgow Coma Scale score 15, and abdominal tenderness to palpation in the left upper quadrant without guarding. Point-of-care clinical ultrasound demonstrated a large amount of free fluid in the splenorenal recess and Morrison's pouch (Figure 1).

Laboratory testing showed hemoglobin 10.3 g/dL; leukocytes, platelets, and coagulation within normal limits; and C-reactive protein 1.3 mg/L. Based on ultrasound findings, abdominal computed tomography (CT) revealed a heterogeneous perisplenic collection (7 × 11 cm) suggestive of hematoma, with abundant free fluid in the perihepatic, paracolic, interloop, and pelvic spaces (Figure 2).

The surgical team was consulted and splenectomy was indicated. Prior to surgery, she developed hypotensive trends (BP, 70/40 mmHg; HR, 106 bpm) with a 2-point drop in hemoglobin compared with initial values, and received 2 units of packed red blood cells. Intraoperatively, a large hemoperitoneum (1,500 cc), perisplenic hematoma, upper-pole laceration, and splenic decapsulation were identified. Surgical and postoperative outcomes were favorable. Histopathology of the specimen showed focal subcapsular intraparenchymal hemorrhage, with no additional diseae. Serologic testing was negative.

The pathologic mechanisms leading to splenic rupture are multifactorial.[5] Most atraumatic ruptures involve an underlying etiologic factor.[1] Causes include hematologic diseases such as chronic myeloid leukemia,[3] infectious diseases including influenza, SARS-CoV-2, or *Streptococcus pneumoniae*,[2] and malignancies. Less frequent causes include drugs (granulocyte colony-stimulating factors, melphalan, anticoagulants, or thrombolytics), chemotherapy, mechanical disorders, and, in a minority of cases, patients without underlying disease.[3]

Atraumatic ruptures may present in a single stage—rapid evolution to hypovolemic shock—or in 2 stages, initially forming a subcapsular hematoma that presents subacutely with epigastric or left upper quadrant pain radiating to the left shoulder (Kehr's sign) due to diaphragmatic irritation, followed by delayed capsular rupture.[1]

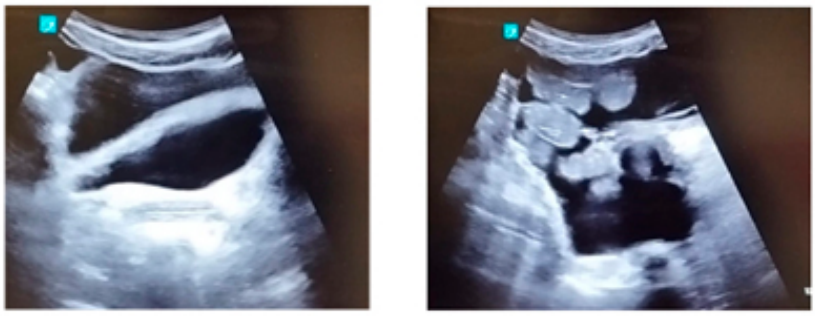

Figure 1. Abdominal point-of-care ultrasound showing intraperitoneal free fluid.

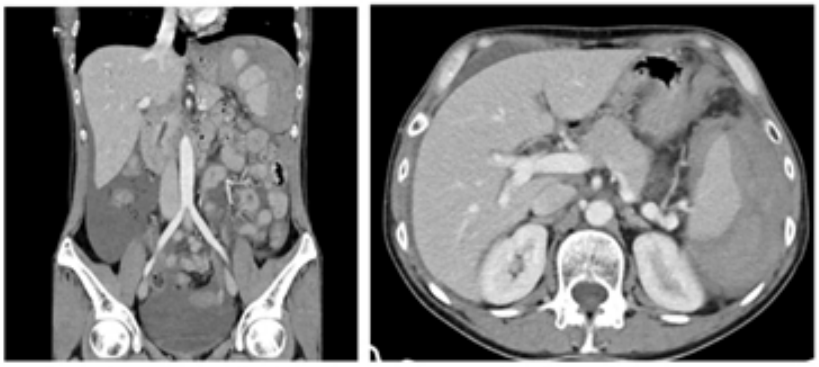

Figure 2. Abdominal computed tomography demonstrating splenic hematoma and free fluid.

Mortality rate is approximately 12%,[3] making it an emergency that requires early treatment to ensure survival; early detection is essential.[1]

Point-of-care ultrasound has been shown to have high sensitivity and specificity for diagnosing severe abdominal pathology,[4] including detection of intraperitoneal free fluid, enabling faster, optimized management and shorter time to surgical treatment.[5]

In our patient, the proposed triggering mechanism was an increase in intra-abdominal pressure associated with vomiting.[6] Due to early clinical ultrasound—despite the patient's initially stable hemodynamic state, preserved general condition, and absence of leukocytosis or elevated inflammatory markers—a CT scan was promptly requested before hemodynamic instability developed, allowing rapid surgical intervention and recovery.

For these reasons, point-of-care ultrasound is a technique that every emergency physician should be able to perform[7] and should be available in all emergency departments.[8]

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